

CHAPTER 1

Developments in the Developmental Approach to Intellectual Disability

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DEVELOPMENTS IN THE DEVELOPMENTAL APPROACH TO INTELLECTUAL DISABILITY

The developmental study of intellectual disability is a long-established forerunner of developmental psychopathology

"Make for yourself a teacher, acquire for yourself a friend, and judge everyone in a positive light . . ." Ethics of the Fathers (Pirkei Avot, 1, 6). We dedicate this chapter in honor of Ed Zigler for all his contributions to the science and welfare of so many children, including those with intellectual disability. He is the most inspiring teacher, loyal friend, and positive influence to all who know him. As with so much of his work, his articulation of the developmental approach to intellectual disability helped humanize our understanding of a population that for too long had been underserved and kept at the fringes of society. We are especially grateful to Dante Cicchetti for inviting us to contribute this chapter, on a topic about which we are so passionate and about which he was a visionary. His leadership both in the world of science and

with origins that predate the formal emergence of the latter discipline by decades, and yet is still in its early, and sometimes apparently regressive, stages of developmental emergence relative to other areas of work. It was largely shaped by scholars, such as Heinz Werner, Edward Zigler, and Dante Cicchetti, who were also seminal to the emergence of the scholarly discipline of developmental psychopathology, and yet it is often conceptualized as a separate unrelated entity. These complex relationships provide a lens through which we can understand the advances, and setbacks, in the study of intellectual disability, and its place in the domain of developmental psychopathology

in making the world a better place for children is an example to us all. We thank Jillian Stewart, Johanna Querengesser, Ashley Reynolds, Icoquih Badillo-Amberg, David McNeil, Eric Keskin, and Martina Tiberi as well as other members of the McGill Youth Study Team for their help in preparing the manuscript.

(and thereby in this volume). Within this framework, we highlight the thinking and research that led to and continue to maintain the developmental approach to intellectual disability and consider them with regard to developments in the study and understanding of intellectual disability since Hodapp and Burack's (2006) chapter in the last edition of this handbook (Cicchetti & Cohen, 2006).

As intellectual disability is essentially defined by the development of cognitive abilities and, to a considerably lesser extent, social skills that are so delayed and ultimately impaired that it only involves a small percentage of persons, it is a paradigmatic example of the construct of development at the extreme that is so essential to the field of developmental psychopathology (Burack, 1997; Cicchetti & Pogg-Hesse, 1982). In this way, intellectual disability also exemplifies Urie Bronfenbrenner's notion of an experiment of nature, which could never be replicated in an experimental setting but in this case is informative for understanding the course of typical cognitive and social development. Thus, consistent with Cicchetti's (1984) dictum that "you can learn more about typical functioning by studying its pathology and more about its pathology by studying its typical state" (p. 4), intellectual disability is a window into addressing issues and questions about cognitive and social development that cannot be fully answered when only considering typically developing persons (Burack, 1997; Cicchetti & Pogg-Hesse, 1982; Hodapp, Burack, & Zigler, 1990). In providing the example of extreme delay, or impairment, intellectual disability would appear to allow us the opportunity to examine the integrity of the developmental system from the unique perspective of especially slowed or delayed development (Cicchetti & Beegly, 1990; Hodapp & Burack, 1990; Hodapp & Zigler, 1990). As is often the case in nature, this "slow motion" analysis of cognitive and social development allows for a particularly intense level of scrutiny that cannot be attained with events occurring at their typical speed.

The converse of Cicchetti's dictum is also particularly relevant to the study of intellectual disability as the theories and methodologies that have governed the study of development among typically developing persons have, during the past half century, transformed the way that persons with intellectual disability and their families are studied, understood, educated, and supported (for related collections, see Burack, Hodapp, Iarocci, & Zigler, 2012; Burack, Hodapp, & Zigler, 1998; Cicchetti & Beegly, 1990; Hodapp, Burack, & Zigler, 1990). These advances are the focus of this chapter as we highlight the ongoing and evolving conceptual, methodological, and interpretive contributions of the so-called developmental approach to

the study of persons with intellectual disability and the ways that they have led to a more precise and sophisticated science (Burack, Dawkins, Stewart, Flores, Iarocci, & Russo, 2012; Burack, Russo, Flores, Iarocci, & Zigler, 2012; Cicchetti & Ganiban, 1990; Hodapp, Burack, & Zigler; 1990).

THE DIAGNOSIS OF INTELLECTUAL DISABILITY AND ITS (LACK OF) MEANINGFULNESS

Although the diagnosis has far-reaching implications for the development and outcomes of the affected persons, intellectual disability cannot be considered at all tangible. Virtually unique among the phenomena addressed in this volume, the designation of intellectual disability is essentially based on a behavioral classification culled from scores on single measures used to operationalize each of two constructs—in this case, primarily the construct of intelligence but often also that of social adaptation. Typically, people who score in approximately the lowest 3% of the population, or two or more standard deviations below the mean, on standardized tests of intelligence and behavioral adaptation are considered to be intellectually disabled. However, the utility of this designation is compromised in two essential ways. One, the cutoff score is entirely arbitrary. Two, the low IQ, or behavioral, scores can be attained for different reasons and with different profiles as evidenced by the study of the handful of the most common of the more than 1,000 possible etiologies, each of which seems to differ considerably from the other conditions and situations associated with intellectual disability (for reviews, see Burack, 1990; Burack, Hodapp, & Zigler, 1988, 1990; Cornish & Wilding, 2010; Dykens, Hodapp, & Finucane, 2001). These group differences are especially apparent in developmental rates and trajectories as well as the profiles of relative strengths and weaknesses across the myriad of cognitive and social skills that are thought to impact intelligence and the many sub- and sub-sub-tests that make up the various different IQ tests and indexes of behavioral adaptation that are used for the diagnosis. Even given the usual within-group differences that are found in any population, the compelling and clearly demarcated group discrepancies on many aspects of functioning highlight the profound and complex developmental effects of the genes–brain–behavior relations associated with each syndrome that virtually swamp any generalized developmental effects of simply lower levels of intelligence and social adaptation (for relevant collections, see Burack et al.,

1998; Burack, Iarocci, et al., 2012; Howlin, Charman, & Ghaziuddin, 2011; Tager-Flusberg, 1999).

With these pervasive group differences across virtually all aspects of being and functioning, the notion of a population of persons with intellectual disability is a mirage. Thus, the phenomenon precludes a science or study of intellectual disability per se but rather would appear to necessitate the invocation of multiple sciences of identifiable populations that differ meaningfully from each other with regard to etiology, defining features, and developmental trajectories. In this framework, both the concept and the field of intellectual disability are inherently deconstructed from the monolithic framework of a single problem and population to more precise, albeit with the consequent of increasingly complex, conceptualizations and research (Burack, Russo, et al., 2012). This demise of a single science or framework of intellectual disability would seem to come at considerable cost. Prior so-called knowledge about persons with intellectual disability as a whole needs to be forsaken, while the alternative of the imposition of multiple fine-grained fields of study based on clearly differentiated populations inevitably entails considerably more work and would seem to signal the abandonment of any sense, or even hope, of a comprehensive and cohesive field of study. Yet, when the additional work is associated with the imposition of a developmental approach that is premised on the notion of a systemic, organized, and universal system, it provides both more precise information and a unifying framework with which the various fragments of information can be grouped into a meaningful area of scholarship (Burack, 1997; Burack, Iarocci, et al., 2012; Cicchetti & Ganiban, 1990; Cicchetti & Pogge-Hesse, 1982; Karmiloff-Smith, 2009; Zigler, 1969, 1973; Zigler & Balla, 1982; Zigler & Hodapp, 1986).

Diagnostic Criteria and Assessment

In one sense, intellectual disability is relatively easily conceptualized. Across the decades and even centuries of work with persons with intellectual disability, the basic notion has been that a certain percentage of persons function at such low levels of intelligence and social adaptation that they cannot survive or function independently in society, or at least need some intensive external support to do so (e.g., Luckasson et al., 2002; AAIDD, 2010; for reviews, see Rosen, Clark, & Kivitz, 1976; Zigler & Hodapp, 1986). However, intellectual disability is also an odd and elusive behavioral classification as it involves the grouping of an extremely heterogeneous population according to amorphous concepts, arbitrary criteria, and prevailing

societal values, all of which have changed often during the past century (for recent discussions, see Bertelli et al., 2014; Salvador-Carulla et al., 2011; Schalock et al., 2010; Shulman, Flores, Iarocci, & Burack, 2011).

Diagnosis and Classification From a Developmental Perspective

Even as researchers and practitioners of intellectual disability discuss relevant issues, including the meaning of intelligence, the role of indexes of social competence in the classification, the measures that should be used for testing, and specific criteria for a diagnosis, the pragmatic reality is that the classification of intellectual disability has historically been entirely determined by IQ, a quantitative measure of the development of the elusive construct of intelligence. Despite the many concerns about IQ scores, they continue to be essential in the demarcation of persons with intellectual disability and in other strategies to identify persons at developmental risk because IQ scores in childhood are seen as reliable indexes of both a child's current rate of intellectual development and of future levels of functioning in relation to one's same-aged peers (Shulman et al., 2011).

The relative reliability among IQ scores at different points in the lifespan captures the developmental essence of intelligence testing and scores. Although IQ scores are typically discussed as measures of individuals' intellectual abilities in relation to those of other persons of the same age, they are historically and essentially indicators of the rate of development of intellectual abilities that is considered both to be consistent at least through adolescence and to be associated with relative level of functioning in adulthood. The developmental nature of IQ is evident in its formula, which involves dividing mental age (MA), the level of cognitive development attained by the individual, by *chronological age* (CA) and then multiplying that number by a constant (usually 100) (for discussions, see Hodapp, Burack, & Zigler, 1990; Zigler & Hodapp, 1986; Shulman et al., 2011). This is fundamentally a measurement of rate as, in this context, CA (the denominator) represents the amount of time taken to attain the level of abilities indicated by MA (the numerator). As a quick analogy, one might think of a common measurement of rate in our everyday lives, that of speed for which we commonly use the equation of kilometers (or miles) per hour. In this equation, the numerator is the specific distance travelled as indicated by the number of kilometers and the denominator is the length of time, as indicated by the number of hours, which has passed during the travels. Both the numerator and denominator grow in relation to each

other, although the ratio between them varies considerably as it serves both as an indicator of the speed of travel and as a point of comparison. And by definition of rate, faster speeds are associated with ratios in favor of the numerator over the denominator. For IQ, the numerator MA is the developmental distance traveled by the individual in their attainment of cognitive abilities and the denominator CA is the length of time that the individual has lived. In this case, a greater numerator, or more distance travelled, in relation to a smaller denominator, less time passed, indicates faster developmental growth. Clearly a child who attains the types of skills common to a 6-year-old in 6 years, and therefore has an IQ of 100 according to our developmental formula, is developing faster than a child for whom it takes 8 years and has an IQ of 75 but slower than one for whom it took only 5 years and who has an IQ of 120.

In terms of long-term consequences, these rates of development are highly correlated with IQ scores and related levels of functioning throughout the rest of the lifespan. Thus, slower development and, therefore, lower IQ scores in childhood are inevitably associated with lower scores on IQ tests and generally lower levels of functioning in adulthood. IQ scores are typically standardized in that they are normed across a representational sample, and, thereby reflect a statistically calculated average for a specific level of functioning at a given age or period in life. Within this context, the primary criterion for the designation of intellectual disability generally involves a cutoff score on a standardized IQ test that is associated with some statistical designation. This cutoff score is usually 70, which is two standard deviations below the mean for the general population for which a mean of 100 is statistically designated. According to this statistical designation, persons with intellectual disability should represent approximately the bottom 3% of the population in terms of IQ scores, although the number of persons who are actually diagnosed often varies in relation to income level of country (Carulla et al., 2011), age of the individual (Hodapp & Zigler, 1986), and the use of social adaptation criteria in addition to the IQ score for the diagnosis (MacMillan, Gresham, & Siperstein, 1993; Switzky & Greenspan, 2006).

The Arbitrary and Amorphous Nature of the Criteria for the Diagnosis of Intellectual Disability

This variability in the specific number of persons who meet this criterion might raise some concerns about the application of two standard deviations below the mean IQ as the cutoff point for the diagnosis, except for the fact that this designation is entirely arbitrary and has no

inherent scientific significance with regard to differentiating functioning level among individuals. Rather, the most meaningful aspect of the score is its statistical convenience as the commonly used nomenclature of standard deviations allows for an easily described percentage of persons. As the standard deviations on common IQ tests represent 15 points, persons with IQs just above (i.e., 71, 72, 73) and below (69, 68, 67) the designated cutoff scores do not differ significantly, either statistically or pragmatically, from each other, whereas the differences among individuals within the range of either intellectual disability, with possible IQs of 0–70, or of so-called typical functioning, with possible IQs of 71 and above, are often vast and can encompass several standard deviations. If that is the case, why are IQ cutoffs imposed? The primary rationale is that some criterion is needed as a standard for social policy and decisions about who should receive specialized services. In this way, the designated score which is based on a statistical model of the real world distribution of IQ scores, and thereby level of functioning, offers a rough estimate of the number of persons who should be and are eligible for some combination of additional funding, resources, services, and supports from governmental and other service agencies. The delineation of intellectual disability then is not a scientific one, but rather one that is largely dependent on societal values and discourse, as well as the resources that are made available to support persons with intellectual disability.

The extent to which the designation of intellectual disability is an amorphous and arbitrary concept is evident in that the specific cutoff and the associated definition has been modified at least nine times over the past 100 years in the United States (AAMR, 2002; Harris, 2005; Zigler & Hodapp, 1986) and is once again revised in the newest versions of both the American-based, Diagnostic and Statistical Manual of the American Psychiatric Association (DSM-5) (APA, 2013), and the International Classification of Diseases (*ICD*) (Bertelli et al., 2014; International Advisory Group for the Revision of *ICD-10* Mental and Behavioural Disorders, 2011; Salvador-Carulla et al., 2011). These changes in both the diagnostic criteria and the nomenclature are due to many different “real” factors including new knowledge regarding the causes and types of intellectual disability, the perceived significance of social competence and adaptive behaviors, perspectives on eventual outcomes and well-being of persons with intellectual disability, attitudes of societies and policy makers toward individuals’ dignity and roles in the community, and pragmatically the resources available for services and intervention. Yet, they also highlight the arbitrariness of

the designation and the influence of factors external to the individuals with the diagnosis.

The most blatant example of the arbitrariness of the criteria was evidenced at the beginning of the 1960s. With the emerging zeitgeist of the time on the promotion of social justice and the desire to improve the lots and lives of those who were deemed less fortunate, the IQ cutoff score was changed from 70, two standard deviations below the mean of 100, to 85, one standard deviation below the mean, by the American Association on Mental Retardation (Heber, 1961). Accordingly, the percentage of persons diagnosed with intellectual disability grew from approximately 2–3% of the population to 16%. In the United States alone, with a population of approximately 200 million at the time, that meant a jump of about 26 million people, from around 6 million to approximately 32 million, who fell within the parameters of a diagnosis of intellectual disability (Zigler & Hodapp, 1986). The absurdity that so many people could go to sleep one night without a diagnosis and wake up the following morning meeting the primary criterion of a person with intellectual disability highlights the obvious arbitrariness in the designation and construct of intellectual disability that precludes any notion of some intrinsic condition that can be meaningfully quantified. These points are even further underscored by the subsequent reversion a dozen years later to the original IQ cutoff (Grossman, 1973), thereby shedding the same approximately 26 million people of their diagnosis—as if in a fairy tale in which they go to sleep one night and are magically relieved of their problem, which, of course, they had not suffered from prior to the change in diagnostic criteria a decade earlier (Zigler & Hodapp, 1986). These changes in the diagnostic criteria were certainly well meaning with the initial change occurring in an era of relative affluence and an emergent emphasis on social change with the intention of enabling services for those persons who typically did not meet the criterion of intellectual disability, but were still considered to be at risk as their intellectual functioning was at the low end of the average range. This course of events provides insight into the difficulties of conceptualizing intellectual disability and the extent to which it must be considered in relation to contemporary societal norms and values.

The Latest Incarnation of the Diagnostic Criteria for Intellectual Disability

In the latest edition of the *Diagnostic and Statistical Manual of Mental Disorders (DSM-5; APA, 2013)*, the term *intellectual disability* (also referred to as intellectual developmental disorder [IDD]), intended to reflect deficits in

cognitive capacity beginning in the developmental period, replaces the term *mental retardation*, which had long been used in classificatory schemes such as those of the American Association on Mental Retardation (AAMR) and the American Psychiatric Association for the *DSM*. This revised terminology brings the *DSM* into alignment with the World Health Organization's *International Classification of Diseases (ICD)* and other professional disciplines (Salvador-Carulla & Bertelli, 2008; Salvador-Carulla et al., 2011). Intellectual disability is also now the term of choice for public policy discourse by influential groups representing people with intellectual disabilities such as the American Association on Intellectual and Developmental Disabilities (AAIDD, formerly the AAMR; Schalock et al., 2011).

In addition to the revisions in the nomenclature, the changes and revisions to the *DSM* with regard to persons with intellectual disability stem from joint efforts of the American Psychological Association and the National Institutes for Mental Health to expand the scientific basis for psychiatric diagnosis and classification (<http://www.dsm5.org/about/Pages/DSMVOverview.aspx>). The revisions in the *DSM-5* are based on a comprehensive review of scientific advances, targeted research analyses, and clinical expertise. For example, one of the primary changes in the *DSM-5* involves a shift from a categorical to a more dimensional system. In the earlier versions of the *DSM*, disorders were described by category, with a specific list of symptoms within each domain (e.g., communication). In this categorical system, a person either displayed a symptom or they didn't, and the presence of a certain number of symptoms was required for a diagnosis. The newly published *DSM-5* involves dimensional assessments that allow clinicians to rate both the presence and the severity of the symptoms, such as very severe, severe, moderate, or mild.

As *DSM-5* is no longer based on a multiaxial classification system, intellectual disability is no longer listed as an Axis II disorder but rather is classified as a neurodevelopmental disorder of brain development. Both *DSM-5* and the AAIDD use the consensus definition of intelligence as a general mental ability that involves reasoning, problem solving, planning, thinking abstractly, comprehending complex ideas, judgment, academic learning, and learning from experience. However, the term “intellectual disability” as used by the AAIDD is a functional disorder, explicitly based on the WHO International Classification of Functioning (ICF). Within this framework, the extent of disability is evaluated within particular contexts for which the focus is on the supports needed to normalize

an individual's life to the extent possible (Schalock, Borthwick-Duffy, Bradley et al., 2010).

In previous versions of the *DSM*, the extent or severity of intellectual impairment was denoted by the levels of mild, moderate, severe, and profound. Whereas these levels of severity will likely remain as central to the new *ICD-11* system, specifiers (i.e., mild, moderate, severe and profound) are used in *DSM-5* instead of subtypes to designate the extent of adaptive dysfunction in academic, social, and practical domains. Within the AAIDD disability model, the focus is on assessment of the supports needed to accommodate to the specific setting (e.g., home, community).

The *DSM* criteria of an IQ score below 70 and low adaptive functioning do not change for the diagnosis of intellectual disability, but the diagnosis involves a greater reliance on adaptive functioning with greater consideration about how intelligence is applied to the functions of everyday life (*DSM-5*). The age of onset is no longer specifically defined as before 18 years and is replaced with onset during the developmental period. However, intellectual disability is distinguished from neurocognitive disorder, which involves a deterioration of cognitive functioning (e.g., dementia) with late onset. In addition, an overall adaptive functioning score of two standard deviations below the mean is no longer necessary, as the criteria is met when at least *one* domain of functioning—conceptual, social, or practical—is sufficiently impaired, based on an assessment of adaptive functioning, that ongoing support is needed. Under exceptional circumstances when standardized assessments cannot be used, as in the case of persons with sensory or physical impairments, a diagnosis of unspecified intellectual disability may be applied to individuals with significant adaptive functioning impairments (APA, 2013).

Even as the revisions to the *DSM* were welcomed by some stakeholders, they elicited criticisms from others. For example, the AAIDD raised the concern in an open letter to the president of the American Psychiatric Association (<http://aaid.org/docs/default-source/comments/aaid-dsm5-comment-letter.pdf>) that the new proposed changes to the criteria for intellectual disability would cause confusion, suggesting that the new boundaries are not as clear as in the *DSM-IV*. For example, the change in the criteria in *DSM* from necessitating that the IDD be present before 18 years of age, to the proposed revisions that all symptoms must have an onset during the developmental period, leaves the age at which the developmental period ends open to interpretation. The AAIDD suggests that this will result in the inconsistent use of developmental

periods across jurisdictions. It also took issue with the use of the terms adaptive behavior and adaptive functioning as potentially confusing. According to the AAIDD, adaptive behaviors are the conceptual, social, and practical skills a person may have, whereas adaptive functioning is how well, or independently, the person is able to use those skills to handle common demands in life. Thus, in an appeal to John M. Oldham, the president of the *DSM-5*, the AAIDD argued that the *DSM-5*'s substituting of adaptive *functioning* for adaptive *behaviors* as “communication, social participation, functioning at school or at work, or personal independence at home or in community settings” was not consistent with the AAIDD position nor with the current psychometric literature, and was problematic because it did not adequately capture functioning but rather emphasized the individual's skills (<http://aaid.org/docs/default-source/comments/aaid-dsm5-comment-letter.pdf>). Similarly, the changes in the mutiaxial diagnosis also posed a concern for clinicians. For example, Harris (2013) highlighted the high prevalence of psychiatric disorders in people with a diagnosis of intellectual disability and suggested that both diagnoses are often warranted. However, the elimination of the mutiaxial classification and removal of Axis II in *DSM-5* may result in clinicians overlooking diagnoses of mental health disorders, such as anxiety disorder or depression, among persons with intellectual disability.

The Application of the Diagnostic Criteria

The continual fine-tuning of the diagnostic criteria and system represents evolving synergy of professional and societal influences on the ever-changing ways that persons with intellectual disability are viewed, understood, and supported by those around them. As certain problems are addressed, other limitations in the process continue to be identified (Charman, Hood, & Howlin, 2008). For example, even into the twenty-first century, disproportionate numbers of minority children were still being diagnosed with mild intellectual disability as intelligence tests often underestimate the abilities of these children, who also typically fail to meet the dominant culture's expectations concerning academic performance (Hays, 2001; Valencia and Suzuki, 2001). In addition, other non-cognitive factors, such as child's health history, physical impairments, motivation levels, and social milieu must be considered when assessing intellectual abilities as they may inhibit performance on IQ tests or other cognitive tasks, especially among children from atypical or non-majority backgrounds. These types of concerns regarding the narrow focus of, and potential external influences on, intelligence

tests and their use as an exclusive vehicle for defining intellectual disability was the motivating reason for the initial insistence of the American Association of Mental Deficiency (AAMD) (later the AAMR and AAIDD) that low scores on adaptive behavior be included, along with low IQ scores, in its definition of intellectual disability (Borthwick-Duffy, 2007). Accordingly, traditional methods of examining cognitive and adaptive abilities are now supplemented with a thorough examination of the child's developmental history, family history, and social and cultural environment. A broader picture of the child's developmental status is obtained with parent and teacher interviews and evaluations of medical and school records. For example, information about the child's physical problems, peer interactions, social skills and emotional state can be productively used in conjunction with more formal assessment methods, despite their somewhat limited reliability and validity. Consequently, within the framework of a multifactorial model of assessment, the evaluation of intellectual disability is increasingly taking place within an interdisciplinary social developmental framework.

The emphasis is on providing a diagnosis as early as possible to identify the child's specific needs, to ascertain the required services, and to facilitate communication across the team of relevant professionals and among family members. However, this process is not without its problems especially as it can also stigmatize the child if it leads to unwarranted negative projections by professionals and parents about the child's potential. Thus, professionals need to be sensitive to the effects and limitations of the label and recognize that intellectual disability only captures specific aspects of the individual. As the manifestation and outcomes of the key symptoms of intellectual disability vary across the lifespan (Zigler & Hodapp, 1986), a comprehensive assessment program should be used to follow the individual through many of the key periods of development. Current social and behavioral models are characterized by an emphasis on the need for repeated assessments to evaluate the child's changing developmental status and needs, the impact of medical treatments, the appropriateness of educational placements, and the effectiveness of educational programming. This type of approach to assessment captures the dynamic and changing quality of development as well as its social nature. In addition to their discrepancies in cognitive functioning, persons with intellectual disability also differ with regard to their physical, social, and emotional characteristics as well as in their relations with their families and the broader environment.

THE ORIGINS OF THE DEVELOPMENTAL APPROACH TO THE STUDY OF INTELLECTUAL DISABILITY

As with virtually any aspect of scholarship, the developmental approach to intellectual disability has neither a clearly identifiable start date nor even a universally accepted origin. Thus, in a somewhat, but certainly not entirely, arbitrary fashion, we choose an auspicious event that both provided a conceptual link to the essential scholarly antecedents to the field and generated a foundational and vital body of research that transformed the research on persons with intellectual disability by grounding it in developmental theory and methodology. The event was Edward Zigler's 1967 publication of a conceptual piece, "Familial Mental Retardation: A Continuing Dilemma," which appeared in the prestigious general scientific journal, *Science*, at a time when the study of intellectual disability (or mental retardation as it was called at the time) was rapidly growing as a distinct, but largely circumscribed, field. Zigler used that article and a soon-to-follow sister article (Zigler, 1969), "Developmental Versus Difference Theories of Mental Retardation and the Problem of Motivation," which was published in the *American Journal of Mental Deficiency* as forums to challenge the prevailing (so-called) *defect approach* that characterized virtually all of the other research on intellectual disability at the time and continued to influence the field significantly into the 1980s and to some extent even until today (for a collection of pieces on the defect theories and early statements of the developmental approach, see Zigler & Balla, 1982).

The defect approach was largely characterized by a monolithic framework in which intellectual disability was seen as a single entity that was essentially caused by one or more of any number of proposed defects that were common to all persons with the diagnosis. Typically in this approach, cognitive and neuro-cognitive functions or abilities were the primary focus as the empirical work in the field was largely 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 to be essential across all domains of functioning, including cognitive rigidity, memory processes, discrimination learning, and attention-retention capabilities, among many others (for reviews of these approaches and their inherent problems from a developmental perspective, see Burack, 1990; Zigler, 1967, 1969; Zigler & Balla, 1982). With the use of experimental paradigms that were sophisticated for the time, researchers presented evidence

of deficient performance in virtually all of these areas of functioning, and each specific defect was touted as the central cause of intellectual disability. Unfortunately, the evidence was consistently *fatally* flawed as the researchers failed to consider essential and seemingly obvious conceptual and methodological issues, such as the inherent differences in developmental level of functioning between persons with and without intellectual disability of the same CA, the multiplicity of etiologies associated with intellectual disability, the uniqueness of each etiology with regard to phenotypic expression, and social factors related to the life experiences of persons with intellectual disability that could affect performance on experimental tasks (for reviews, see Burack et al., 2001; Burack, Dawkins, et al., 2012; Zigler, 1970, 1973; Zigler & Balla, 1982).

In critiquing and debunking the various claims of the defect theorists, Zigler and colleagues (e.g., Hodapp, Burack, & Zigler, 1990; Zigler & Balla, 1982; Zigler & Hodapp, 1986; Zigler, 1967, 1969) highlighted the conceptual and methodological concerns as the hallmarks of a nascent approach to intellectual disability that would be based on classic developmental theory. The fundamental principles of what would become known as the developmental approach to intellectual disability were based primarily in the previous few decades of developmental theorizing by Heinz Werner and Jean Piaget, but also in two centuries of relevant history of medical scholarship, more contemporary interpretations of statistical analyses of the plotting of IQ scores, and the social-political zeitgeist of the era. In their original and subsequent iterations over the past half-century, Zigler and others, including especially Dante Cicchetti, John Weisz, and Robert Hodapp, contributed immensely to a science and transformative understanding of intellectual disability that were both more precise with regard to the development and functioning of each individual and more humanistic in that they were framed within notions of development that are universal to all persons.

In his initial seminal papers, Zigler (1967, 1969) proposed four principles that both challenged the dogma of the time regarding persons with intellectual disability in one or more ways and that continue to be essential guidelines for current work almost a half century later, albeit in more nuanced and fine-tuned ways of thinking that have developed in the interim since their original articulation. One, the notion that functioning, development, and developmental trajectories differed according to etiology was consistent with centuries of evidence of meaningful differences in behavior in relation to the source (etiology) of the intellectual disability, but challenged the

monolithic framework of intellectual disability as a specific disorder and of the population of persons with intellectual disability as a single population that prevailed in the 1960s–1980s and continues to be at the heart of even some contemporary research. Despite resistance from leaders in the field to any type of etiological differentiation (for a review of examples of the opposition to this innovation by leading scholars in the field, see Burack, 1990), Zigler's (1967, 1969) two-group approach involved the delineation between persons whose intellectual disability is due to familial factors such as genetic transmission of intelligence and those for whom it is due to some type of organic insult. This dichotomy would presage a multiple group, or even a 1,000+ group, approach fueled by evidence of differences in development across virtually all aspects of functioning (i.e., cognitive, language, social, emotional) (e.g., Burack 1990; Burack, Hodapp, & Zigler, 1988, 1990; Burack, Russo, et al., 2012; Dykens, Hodapp, & Finucane, 2000; Howlin, Charman, & Ghaziuddin, 2011; Tager-Flusberg, 1999). The need for this type of precision continues to grow as evolving technology continues to enhance our abilities to identify, highlight, and compare sub and sub-sub groupings of specific etiological groups (e.g., Romano et al., 2014; Dimitropoulos, Ferranti, & Lemler, 2013) as well as sub- and sub-subcomponents of the various domains of everyday functioning (e.g., attention, working memory, language, social skills) (Iarocci, Porporino, Enns, & Burack, 2012; Jarrold & Brock, 2012; Russo, Dawkins, Huizinga, & Burack, 2012; Vicari, 2012).

Two, grounded in the developmental theories of Jean Piaget (1970) and Heinz Werner (1948, 1957), Zigler's (1967, 1969) adherence to the traditional developmental principles of an organized, coherent, and systemic organism led to the delineation of universal patterns of developmental sequences and structures among persons with familial intellectual disability, those whose low IQ scores seem to arise from the genetic transmission of intelligence rather than any type of physiological or organic "insult" (for a discussion, see Zigler & Hodapp, 1986). According to Zigler, development, at least in this group which represents essentially the "purest" form of intellectual disability that is unaffected by specific organic insult, unfolds in a typical way but at a slower rate and with a lower asymptote. This view that the integrity of development is maintained even in the face of extreme delay contrasted markedly from the difference or defect approach in which persons with intellectual disability were discussed virtually always within the framework of differences rather than of similarities or universalities in relation to others. In the two decades following Zigler's initial articulation of the

developmental approach, the notion of intact but delayed development would be tested extensively by Weisz and colleagues (Weisz, 1990; Weiss, Weisz, & Bromfield, 1986; Weisz & Zigler, 1979) and extended conceptually to include persons with Down syndrome by Cicchetti and colleagues (Cicchetti & Beeghly, 1990; Cicchetti & Pogge-Hesse, 1982; Cicchetti & Sroufe, 1976, 1978; Wagner, Ganiban, & Cicchetti, 1990). This latter extension to include persons with Down syndrome within a developmental framework revolutionized the developmental approach to intellectual disability by the de facto inclusion of all persons with some type of organic etiology, whose impairments and profiles of functioning seem qualitatively different from the typical population. In an era of considerable debate and discussion about the integrity and legitimacy of developmental theory (for a discussion, see Bronfenbrenner, Kessel, Kessen, & White, 1986), it also would lead to an essential expansion of developmental theory to a more liberal approach that would include more variability, at least in terms of group differences, than was considered in the more conservative classic developmental approaches articulated by Zigler (Cicchetti & Pogge-Hesse, 1982; Cicchetti & Ganiban, 1990; Hodapp, Burack, & Zigler, 1990). This contribution can also be seen as essential to a more nuanced understanding of the extent to which various aspects of development are related to each other in an inherent versus a happenstance manner (Hodapp & Burack, 1990, 2006) and as the precursor to neuroconstructivism (Karmiloff-Smith, 1998, 2009) and other approaches focused on the trajectory of gene–brain–behavior relationships within specific etiological groups (for examples regarding Williams syndrome, see Elsabbagh & Karmiloff-Smith, 2012; Landau, 2012; for an example regarding *fragile X*, see Cornish, Bertone, Kogan, & Scerif, 2012).

Three, Zigler's emphasis on mental, rather than chronological, age in experimental and observational comparisons between persons with and without intellectual disability led to essential changes in the empirical study of intellectual disability with more fine-tuned questions regarding group differences in specific areas of functioning that could not simply be attributed to more general a priori differences in functioning between the groups. In noting that the finding that persons with intellectual disability perform worse than typically developing individuals of the same CA is circular, Zigler (1967, 1969) highlighted the futility and obvious methodological flaws of the defect approaches that unfortunately persist to some extent even until today. These approaches were derived largely from the inevitable finding of deficits among persons with intellectual disability who were compared with typically

developing persons of the same CA—findings that simply confirmed the obvious conclusion that lower functioning persons perform at lower levels than higher functioning people. With the increased influence of developmental considerations, the matching of persons with and without intellectual disability has become increasingly fine-tuned as groups are often now matched on a measure related to the domain of functioning being studied, thereby further diminishing the possibility that findings of differences or similarities are a confound of the a priori relative strengths or weaknesses associated with specific etiological groups (for relevant discussions, see Burack, 1997, Burack et al., 2012; Burack Iarocci, Flanagan, & Bowler, 2004; Jarrold & Brock, 2004; Karmiloff-Smith, 2009).

Four, the inclusion of the social-motivational personality factors served to emphasize that the understanding of persons with intellectual disability needed to involve the whole person. Zigler (1967, 1969) highlighted that the life experiences of failure, exclusion, and segregation that are common to persons with intellectual disability (as well as to other commonly marginalized populations) have profound effects on each individual. Zigler even argued that apparent deficits found among persons with familial intellectual disability as compared with typically developing persons of the same MA could be the consequences of responding styles adopted by persons with intellectual disability because of their experiences (for reviews, see Merighi, Edison, & Zigler, 1990; Zigler & Bennett-Gates, 1999). These outcomes were most apparent among persons with intellectual disability who were institutionalized, a far more common occurrence when Zigler was writing than it is today although the implications of segregation and exclusion are still pressing issues.

These four transformative principles of the developmental approach, as initially articulated by Zigler (1967, 1970, 1973) and as developed over the years by him and others, comprise the guiding framework for our discussion both of persons with intellectual disability and of the science in which they are studied. We use these principles of the developmental approach as a framework for interpreting both essential historical and contemporary issues in the study of intellectual disability, and highlight ways that the approach has itself developed since its original articulation a half century ago to provide an increasingly fine-tuned and sophisticated science. Conversely, consistent with Cicchetti's (1984) premise that typical and atypical development are mutually informative, the challenges faced in the application of developmental principles to intellectual disability provide insight into essential ways that developmental theory can be adapted (Burack, 1997;

Cicchetti & Ganiban, 1990; Cicchetti & Pogge-Hesse, 1982; Hodapp & Burack, 1990).

THE TWO-GROUP APPROACH AND BEYOND

Zigler's (1967, 1969) original delineation of the two-group approach was ironic in that it hearkened back to even more precise medical and social classifications of intellectual disability that predated it by decades and even centuries, while challenging a contemporary field that was considerably less fine-tuned. Although Zigler's dichotomy between familial and organic intellectual disability was an essential enabler of the deconstruction of the field of intellectual disability, it can be seen as somewhat of a retreat from more fine-tuned, albeit not always accurate, diagnostic and classificatory schemes dating back centuries (for reviews, see Burack, 1990; Scheerenberger, 1982). As early as the end of the sixteenth and beginning of the seventeenth century, the Swiss physician Felix Platter identified two broad groupings of persons with intellectual disability. He described one group of individuals as simple-minded since infancy and the second as those persons born with identifiable physical anomalies that were the manifestations of underlying organic disorders that were the source of the intellectual disability. This differentiation between persons with intellectual disability born with and without observable physical and physiological problems continued to be highlighted in even more precise classification systems by later medical workers interested in intellectual disability. For example, in the latter part of the nineteenth century, both John Langdon Down (1887) and William Wetherpoon Ireland (1877) provided detailed early classification systems in which they distinguished between intellectual disability which appeared to be the consequence of some combination of familial, genetic, environmental, societal, and cultural factors and that which was the outcome of more obvious genetic anomalies or other types of organic insult. Down classified persons with intellectual disability into three general etiological groupings, which he referred to as congenital, accidental, and developmental. Ireland delineated ten categories, nine of which were associated with medical conditions that were linked with intellectual disability, whereas the tenth was labeled as idiocy by deprivation.

In the subsequent classification systems of the twentieth and twenty-first centuries, the number of genetic or organic conditions associated with neurological problems and intellectual disability grew rapidly as a result of increasingly sophisticated technologies and scientific advances.

Current estimates include more than 1,000 organic conditions associated with intellectual disability (Hodapp & Burack, 2006), although the meaningfulness of that number is diminished by the realization that current technology increasingly allows us to identify the substantial effects on performance of even relatively subtle neurological or physiological differences across even persons with the same syndrome (e.g., for a discussion of within-group differences among persons with *fragile X*, see Romano et al., 2014; for a discussion of genetic subtype differences among persons with Prader-Willi syndrome, see Dimitropoulos, Ferranti, & Lemler, 2013).

Zigler's Emphasis on Familial Intellectual Disability

In delineating the two-group approach, Zigler's primary interest as a developmentalist was with the persons with intellectual disability, especially in the mild to moderate range, whose etiology could be considered cultural-familial (for the rest of the chapter we refer to this grouping as familial)—a classification conceptually similar to Platter's simple-minded from birth, Down's developmental in nature, and Ireland's idiocy by deprivation (for twentieth-century systems of classification by etiology, also see Kephart & Strauss, 1940; Lewis, 1933). Consistent with these earlier frameworks, Zigler argued that some combination of familial-genetic and environmental factors affect the development of intelligence and the occurrence of intellectual disability among this group (Hodapp & Dykens, 2001; Zigler & Hodapp, 1986). As predicted by Zigler and Hodapp, current conceptualizations provide increasingly complex but precise frameworks of the genetic transmission of intelligence and its relation to the environment in general and especially in relation to the occurrence of familial intellectual disability (for discussions, see Iarocci & Petrill, 2012; Shulman et al., 2011).

Persons with familial intellectual disability are characterized by IQ scores that are typically in the mild, or sometimes high moderate, range, as they represent a statistically expected downward extension of the typical IQ range and can be thought of as those who were simply at the low end of the normal distribution of IQ (Lewis, 1933; Pearson & Jaederholm, 1914; Penrose, 1963; Zigler, 1967). According to Zigler's conceptualization, familial intellectual disability is simply the case of development that occurs at a slower rate but along the normal continuum of intellectual development. This is consistent with the observation that both individuals with familial intellectual disability and persons with IQs in the lower end of the typical range are likely to have at least one parent with an IQ in or near the range

of intellectual disability (Broman, Nichols, Shaughnessy, & Kennedy, 1987; Hodapp & Burack, 2006; Zigler & Hodapp, 1986). The lack of clear genetic or physiological markers for the children who fit the criterion for familial intellectual disability was underscored a century ago as scores on the early Binet intelligence tests of school children showed neither a natural split nor any significant differences between the children at the lower end of the normal distribution of IQ and a group of typical school children (Pearson & Jaederholm, 1914). This point was reiterated shortly thereafter by Lewis (1933), who argued (in the terms of his time for the group of persons we refer to as those with familial intellectual disability) that there “seems to be a close biological kinship between the subcultural defective and the main body of normal persons . . .” (p. 300), and later by Tarjan (1960), who noted (in the terms of his day for the group of persons we refer to as those with familial intellectual disability) that the “socioculturally deprived” group of persons with intellectual disability blend “gradually into the continuum of the general population” (p. 60; see also Dingman & Tarjan, 1960).

The Politics of Eugenics in Conceptualizing Intellectual Disability Associated With Familial Transmission

The notion of the familial transmission of intelligence has been a historically essential, albeit stigmatizing, aspect of the work with persons with intellectual disability (see Rosen et al., 1976; Siperstein, Norkins, & Mohler, 2007; Zigler & Hodapp, 1986). This focus was highlighted by the advocates of the eugenics movement who, in the late nineteenth century and beginning of the twentieth century, reported on supposed family lines in which feeble-mindedness, criminality, and other forms of behavior that were considered to be generally immoral were linked across generations of families (for a discussion, see Shulman et al., 2011). In one particularly often cited example, Arthur Estabrooks (1916) used the sociologist Robert Dugdale’s (1877; Dugdale, Harris, & Giddings, 1910) report on the criminal history of the so-called “Jukes” family from upstate New York to make the case that the procreation of persons with lower intelligence was detrimental to society. Ironically, Dugdale’s report was originally intended to be used as evidence for the need for better social welfare and improved environments for persons from these backgrounds, but was twisted to support the argument for legal restrictions on their opportunities to reproduce.

The claim that allowing persons with intellectual disability to procreate was detrimental to society was particularly forcefully advocated by the twentieth-century eugenicist Henry Goddard (1912), who argued in his

influential book *The Kallikak Family: A Study in the Heredity of Feeble-Mindedness* for the key role of heredity in feeble-mindedness. In a post hoc recreation of the supposed genetic tree of an American Revolutionary War hero named Martin Kallikak Sr., Goddard compared Kallikak’s descendants from an evidently feeble-minded woman whom he met on his way home from the war and his descendants from his wife who was assumed to be typically intelligent. Goddard claimed that a particularly high percentage of Kallikak’s 480 descendants from the feeble-minded woman could be labeled as socially problematic as he identified that 36 were illegitimate, 33 were sexually immoral, 24 were confirmed alcoholics, three were criminal, three were epileptics, eight kept houses of “ill fame,” 82 died in infancy. In contrast, he claimed that all but three of the 496 descendants from Kallikak’s wife were considered to be intellectually normal, and generally prosperous and morally upstanding. With these supposed differences between these two lines of descendants from Kallikak as his evidence, Goddard concluded that levels of intelligence, sanity, and morality were hereditary, and preached that persons classified as “feeble-minded” should be prohibited from procreating to prevent the spread of intellectual disability and its associated deleterious effects on society. The scientific integrity and merits of Goddard’s work have certainly been questioned over the years. For example, factors other than inherited feeble-mindedness, such as fetal alcohol syndrome, have been suggested as possible sources of the generally deleterious outcome of many of Kallikak’s descendants (Karp et al., 1995). Yet, despite the criticisms of the obviously controversial focus and methodological problems of his work from contemporary perspectives, Goddard’s message of hereditary transmission and intrinsic moral shortcomings of persons with familial intellectual disability was to influence the western world through much of the first half of the twentieth century.

Both Goddard’s and Estabrook’s views on eugenics and intellectual disability are cited as important contributing factors to the passing of sterilization laws in 25 states in the United States between 1907 and 1936 and in the Supreme Court decision that ruled in support of the sterilization laws in Virginia (for a discussion, see Zigler & Hodapp, 1986). Zigler and Hodapp note that the sentiment of the Supreme Court and that of much of the nation at the time was summed up by the eminent Supreme Court justice, Oliver Wendell Holmes Jr., who wrote the majority opinion in support of the forced sterilization of a young woman named Carrie Buck, who was forced by the courts in 1927 to undergo compulsory sterilization because she

was thought to be feeble-minded. In his opinion, Justice Holmes wrote, “It is better for all of the world, if instead of waiting to execute degenerate offspring for crime, or let them starve for their imbecility, society can prevent those who are manifestly unfit from continuing their kind . . . Three generations of imbeciles are enough” (*Buck vs. Bell*, 1927). This pronouncement highlights the extent to which the notions of both the genetic transmission of intelligence and the link between lowered intelligence and criminal behavior were instilled in the American psyche in the first half of the twentieth century. Tragically, these pernicious American laws and attitudes would be copied in other places in the world and have even been cited by some as sources of inspiration for the sterilization and extermination practices of the Nazis toward Jews, Gypsies, homosexuals, and others (Kühl, 1994).

The Familial Inheritance of Intellectual Disability

Based on the notion that persons with familial intellectual disability represent a downward extension of the natural variation in intellectual ability, a variety of possibilities related to the interplay of the genetic transmission and environmental influences on intelligence in this population have been forwarded (for reviews, see Hodapp & Zigler, 1995; Iarocci & Petrill, 2012; Simonoff, Bolton, & Rutter, 1998; Zigler & Hodapp, 1986). These include attempts to fine-tune the understanding of both the genetic process in the transmission of intelligence and the interplay between intellectual heredity and environmental influences.

Consistent with the notion that persons with familial intellectual disability meld into the typically developing population, polygenic inheritance models are highlighted in the explanations of the transmission of intellectual disability within families (for a variety of relevant reviews from the past half century, see Gottesman, 1963; Iarocci & Petrill, 2012; Plomin, 1999; Zigler & Hodapp, 1986). According to this framework, many human traits are continuously distributed and are determined by a number of genes that work independently and additively to produce the particular trait whenever normal environmental conditions prevail. Within the framework of combined genetic and environmental contributions to phenotypic outcome, the primary goal is to identify the polygenes that additively accumulate genetic risk and, together with environmental risk factors, increase the likelihood of familial intellectual disability (Zigler & Hodapp, 1986).

In discussing intelligence in relation to familial intellectual disability, Iarocci and Petrill (2012) likened the nature of intelligence to an elastic band, for which environmental conditions may broaden or constrain inherent potential, but only within the limits of the elasticity, or integrity,

of the structure (i.e., inherited familial genes). This is consistent with decades of evidence supporting the notions of the interplay between genetic and environmental factors in the developmental outcome of familial intellectual disability (Richardson, Koller, & Katz, 1985) as intelligence is classically thought to emerge through the coalescence of inherited developmental structures and the organism’s ongoing and active engagement with the environment (Shulman et al., 2011). This relationship between familial transmission and environmental influence in intellectual disability was specifically examined by Broman et al. (1987) in a longitudinal study of pre- and perinatal causes of intellectual disabilities as they sought to differentiate the developmental trajectories of persons with familial intellectual disability from those with a clear organic etiology. Based on findings of a 12-fold increase in the frequency of intellectual disability among full siblings of children with mild intellectual disability, and significantly more affected relatives than among the siblings of children with severe intellectual disability, Broman et al. concluded that mild, but not severe, intellectual disability shows a familial link with normal variation in general intelligence. When the children were classified according to socioeconomic status (SES), SES level was associated with intellectual disability at 7 years. In the group of children in the bottom 25% of SES, 3.3% scored in the IQ range of intellectual disability, whereas only 1.3% of the children from middle 50% and 0.3% of the children from the highest 25% SES groups scored in that range. This is consistent with other evidence that mild, nonorganic, intellectual disability is found primarily at lower SES levels, whereas the prevalence of moderate to severe intellectual disability (IQ < 50) is generally distributed relatively evenly across SES levels (Tarjan, 1970; Richardson & Koller, 1996).

The clear link between the familial transmission of mild intellectual disability and low SES highlights the complexity of disentangling the relative contributions of the genetic transmission of intelligence and the environmental factors. This transmission of intelligence is not simply the case of the familial passing of genes, but involves a complex relation between the occurrence of low IQ in the family and the environment in which the family lives. For example, children of parents with intellectual disability are more likely to be raised in settings with fewer resources than typically found in contexts that are more conducive to optimal development (Simonoff et al., 1998). This may be particularly exacerbated among minority populations who already may have fewer opportunities to optimize child development because of linguistic, cultural, or other mismatches with the majority culture in educational and other settings. These qualifications

highlight the complexity of understanding either the unique contributions or the confluence of issues related to the genetic heredity of intelligence and environmental effects associated with familial intellectual disability.

Differentiating Among Organic Etiologies: Extending Beyond the Two-Group Approach in the Quest for Increased Precision

Although aware of the several different etiological groups that had been cited in the two to three centuries previously and that advancing technology would help delineate many more, Zigler simply proposed a two-group approach. This approach allowed him to highlight the group of persons with familial intellectual disability for whom he hypothesized that development unfolded in the same way as for typically developing persons, albeit at a slower rate, against the backdrop of persons with organic intellectual disability for whom Zigler argued that specific organic insult precluded any necessity of typical developmental patterns. Whereas familial intellectual disability represented a quantitative difference from so-called typical development because it simply represented the lower end of normal variation in IQ, the organic etiologies associated with intellectual disability reflected a qualitative break due to the physiological anomalies associated with each of them. Although Zigler would eventually collaborate with colleagues both on calls for more precise differentiation among persons with organic etiologies (e.g., Burack, Hodapp, & Zigler, 1988; 1990) and on attempts to apply developmental theory to specific etiological groups (e.g., Hodapp, Burack, & Zigler, 1990; Hodapp & Zigler, 1990), his primary emphasis continued to be on the developmental approach as applicable to persons with familial intellectual disability.

Burack et al.'s (1988; 1990; Burack, 1990) subsequent call for the precise differentiation among organic etiologies hardly seems revelatory from the perspective of today or even from the perspective of the time in which it was written as unique developmental trajectories and characteristics had already been identified, but it was in clear contradiction to the prevailing thinking of the time among the leading researchers of intellectual disability. Even as the defect theorists faded from the forefront of the study of intellectual disability by the 1980s, the prevailing leaders still refused to acknowledge the advantages of studying individual etiological groups (for some quotes by leaders in the field expressing the futility of differentiating by etiology, see Burack, 1990). Goodman (1990) expressed much of the spirit of the time in her argument against Burack et al. (1988) that "it is unhelpful and possibly detrimental to make etiology the cornerstone of a diagnostic system . . ."

(p. 466), and that research and intervention would be better served by a focus on other factors, especially level of intellectual functioning. In the interim of the past quarter century, Burack et al.'s position has clearly been vindicated with the ever increasing precision in delineating both etiologies and unique profiles of functioning that are associated with at least those etiologies that are sufficiently common to be studied.

The contribution of differentiating among organic etiologies to the study of persons with intellectual disability is that of precision. And that quest for precision is characterized by the very undevelopmental process of deconstruction of the population of persons with intellectual disability. This process was central to the de facto breakdown of the field of intellectual disability *per se*, and the advent of essentially a conglomeration of many sciences (Burack, Russo et al., 2012). It involves the bottom-up process of grouping by etiology, in addition to other relevant factors such as age and gender, to better understand the functioning of each individual or group of individuals. Big stories, such as those once promulgated by the defect theorists concerning all persons with intellectual disability, are long abandoned in favor of more precise minutia—the research is focused on increasingly fine-tuned studies of components (and sub- and sub-sub-components) of functioning in specific etiological groups (and sub-groups). These sciences can be framed in at least one of two apparently orthogonal, but also overlapping strategies, with the emphasis on a specific aspect of functioning and the manifestation of its various components across various etiologies (e.g., for reviews on attention, short-term and working memory, executive functions and memory as manifested in different etiological groups, see respectively Iarocci et al., 2012; Jarrold & Brock, 2012; Russo et al., 2012; Vicari, 2012), or with regard to a specific etiological group in relation to their functioning within or across the many different domains, sub-domains, and sub-sub-domains of one or more specific areas (e.g., for extensive reviews of the abilities and impairments of persons with fragile X, Down syndrome, and Williams syndrome in the domain of language, see, respectively, Abbeduto, McGuffie, Brady, & Kover, 2012; Chapman & Kay-Raining Bird, 2012; Mervis, 2012).

The ultimate goals of this approach is to better understand the links among genes (loosely defined), brain functioning, and behavior in the pursuit of providing more information about the specified group, the contributors to specific aspects of functioning, and potential interventions at many different levels. The more precise the better, with improving technology in every aspect of the gene–brain–behavior relationship allowing us to

further deconstruct our understanding of each into even smaller more detailed units or sub-units. As these foci are syndrome-specific (or now even sub-syndrome-specific), the notion of a scientific field of intellectual disability is rendered essentially meaningless. Any attempts to discuss intellectual disability in terms of groups in which persons with different etiologies are included (often referred to as mixed-etiology) are futile as the syndrome-specific patterns offset each other in this hodge-podge, thereby obscuring any meaningful patterns—much like the flavors of the individual ingredients in stew are unrecognizable after they've been cooked together. And, as the flavor of a stew can be altered in relation to the ingredients, so too can the findings vary in relation to the makeup of the etiological mix. Thus, the notion that a so-called mixed etiology group reflects the general population of intellectual disability is fallacious, and the use of these groups as target groups or even as comparison groups must be seen as both study-specific and misleading. Rather, the more instructive approach to furthering our understanding of the significance of the gene–brain–behavior relations in these groups is to extend Cicchetti's (1984) dictum of the reciprocally informative relationship between typical development and atypical development to focus on the reciprocally informative relationship among various types of atypicality, in this case the specific syndromes.

One example of the ways that information from different syndromes can converge to inform both about each other and functioning more generally is exemplified in the study of multiple object tracking (MOT) among persons with Williams syndrome and Down syndrome. According to Pylyshyn (2001), the operations necessary to index and track multiple items at once in multiple object tracking are also necessary for deriving spatial relations. Concordantly, O'Hearn and colleagues (O'Hearn, Hoffman, & Landau, 2010; O'Hearn, Landau, & Hoffman, 2005) found poor tracking performance among persons with Williams syndrome that was consistent with their commonly reported problems on visuospatial tasks. Subsequently, Brodeur, Trick, Flores, Marr, and Burack (2013) hypothesized that if poor MOT performance is always associated with visuospatial deficits, then MOT should be relatively spared among individuals with Down syndrome who display strengths in visuospatial processing relative to other aspects of their cognitive functioning. However, contrary to their predictions, they found that the performance on a MOT task was worse among individuals with Down syndrome as compared with MA-matched typically developing children. In explaining these unexpected findings, Brodeur et al. suggested that tracking deficits may occur for

different reasons in the two groups. For example, compared with individuals with Williams syndrome, persons with Down syndrome may display relatively intact object-based selection, but may nonetheless exhibit reduced tracking performance because the temporal resolution of the selection processes is poor, which would create particular difficulties when processing dynamic displays. Another possibility is that object-based selection may be preserved in persons with Down syndrome, but that limitations in executive function may reduce the ability to store information for later report while updating item positions. These post hoc explanations point to the need to specifically fine-tune our understanding both of the strengths in visuospatial functioning among persons with Down syndrome and of the extent to which MOT is generally associated with visuospatial performance. At a more conceptual level, they also highlight the necessity for greater nuance in understanding the abilities and factors that contribute to performance on tasks within a single etiological group and, more complexly, in comparisons across groups.

In retrospect from an era in which research is increasingly informed and fine-tuned by technology such that more and more groupings and subgroupings are defined and studied, Zigler's (1967, 1969) articulation of the two-group approach can be seen as a catalyst in transforming, or deconstructing, the field of intellectual disability into a more precise, albeit increasingly differentiated, science. As evident in the cutting-edge work by Romano et al. (2014) in delineating two biologically and clinically distinct phenotypic outcomes of fragile X using topological data analysis of variations in brain structure and by Dimitropoulos et al. (2013) in identifying differences in language functioning between the genetic subtypes of Prader-Willi, the level of precision and differentiation in the field will continue to increase dramatically. However, as discussed in the next section, Zigler and, to an even greater extent, Cicchetti provide a framework for the integration of these seemingly impossibly fractionated pieces—the fulfillment of Werner's orthogentic principle.

APPLYING DEVELOPMENTAL PRINCIPLES TO THE STUDY OF PERSONS WITH INTELLECTUAL DISABILITY: CLASSIC AND EXPANDED VERSIONS

Zigler and the Classic Developmental Approach

The defining developmental innovation in Zigler's approach to the study of intellectual disability was his imposition of essential classic developmental principles

to the study of persons with intellectual disability, who had previously virtually always been considered defective, impaired, or at least different in some specific way. In citing the organismic-developmental theories of Heinz Werner and Jean Piaget, Zigler (1967, 1969) invoked the notion of universality of the human organism, thereby including the previously excluded group of persons with intellectual disability, at least those with the familial form. Conversely, this approach was also essential to extending classic developmental theory to persons with intellectual disability and, thereby, to provide much needed tests of the theory from the context of atypical development. Even as classic developmental theory, in general, and Piagetian notions, in particular, were under siege throughout the 1960s and beyond as post- and neo- Piagetian scholars, among others, decried the lack of consideration of group and individual differences and even suggested the end of the age of development (Kessen, 1984), Zigler's framework provided compelling, if not sufficient, tests of the universality of classic developmental theory.

Zigler framed his notion of the applicability of the universality of development largely within the context of Werner's (1957) organizational principle of development that "wherever there is life there is growth and development, that is, formation in systematic, orderly sequence" (Werner, 1957, p. 125) and the Piagetian notion of universal sequences of development. These notions resonated especially with Zigler with regard to the population of persons with familial intellectual disability whose lower IQs represent simply a statistically expected downward extension of the typical IQ range as is consistent with the case of any hereditarily transmitted trait (e.g., height, weight). In contrast to the qualitative break that he suggested might be the case with persons with an organic etiology, Zigler hypothesized that the patterns of development of persons with familial intellectual disability should be indistinguishable from those of typically developing persons but that they would unfold at a slower rate and ultimately plateau below the general population mean. Zigler's approach led to two hypotheses of basic developmental similarities, similar-sequence and similar-structure, between persons with intellectual disability and typically developing persons that would be tested extensively by John Weisz and colleagues (e.g., Weiss, Weisz, & Bromfield, 1986; Weisz, 1990; Weisz & Yeates, 1981; Weisz, Yeates, & Zigler, 1982; Weisz & Zigler, 1979).

The Similar-Sequence Hypothesis

The similar-sequence hypothesis reflects the basic tenet of developmental theory that the development of a specific

area of cognitive functioning necessarily follows a clear and universally prescribed order of attainment of the relevant abilities (Hodapp, 1990). The sequences can be thought of as the vertical aspects of development since they involve the trajectory of development in relation to the attainment of one or more specific functions, or abilities, over time. In an extensive review of relevant studies, Weisz and Zigler (1979) found that the integrity of typical Piagetian sequences seems well entrenched among all groups of persons with intellectual disability as it is among virtually all other groups of persons (for discussions, see Weisz, 1990; Hodapp & Burack, 2006).

In considering the inevitability of the universality of Piagetian and other developmental sequences, Hodapp (1990) proposed four possible reasons for their universality. One was that psychological, like biological, development involves a process that is consistent with Werner's (1957) orthogenetic principle, in which "wherever development occurs it proceeds from a state of relative globality and lack of differentiation to a state of increasing differentiation, articulation, and hierarchic integration" (p. 126). The case of human fetal development is a classic case in the realm of biological growth as we all begin as a speck of a single cell that lacks any identifiable form, but unfolds with increasingly identifiable internal and external parts that continue to evolve into a complex human being in which all the parts are distinct and at the same time integrated in that they function in an organized coherent way. In an example of the orthogenetic principle in psychological development, children first use only sentences made of single words to communicate, but further along in development use more complex sentences composed of multiple different words combined in infinite combinations, giving rise to more complex communication. As psychological development is constrained and dependent on biological development that is universal, Hodapp suggests that it too must be universal. Hodapp's second argument is based in the logic that since behaviors are sequentially embedded in each other, a child is only able to perform a difficult or complex task by first attaining the necessary skills through the performance of more basic (earlier) tasks. Hodapp's third argument is based on universally shared information-processing capacities that are used to understand and experience the laws and relationships in nature that transcend culture, geography, or language. Hodapp's fourth argument is that the essential task of all cultures is to foster the development of its children and that this quest may involve universal aspects of the environment, regardless of culture or context.

The Similar-Structure Hypothesis

In contrast to the verticality of his similar-sequences hypothesis, Zigler's (1967, 1969) similar-structure hypothesis is relevant to the horizontal dimension of development as it concerns the relations across domains, or areas, of functioning at one or more given points in time. It is based on the classic developmental principle of an organized system, such as espoused by Piaget in his notions of cognitive structure and stage theory, in which aspects of development are meaningfully interrelated and therefore, despite individual variation, consistently emerge and unfold in some universally systematic and hierarchic way with consistent temporal patterns (also see Werner, 1957). In this manner, developmental profiles across domains should, with only minor differences, be consistent across the population of typically developing individuals for any given point in time (age) of development. Thus, also unlike the similar-sequence hypothesis, the similar-structure hypothesis was considered by Zigler to be specific to persons with familial intellectual disability for whom he considered the patterns of development to be the same as in typical development, but to just unfold at a slower rate and to ultimately reach a lower plateau—delayed rather than different (Zigler & Hodapp, 1986). In direct contradiction to the prevailing difference or defect theories of intellectual disability of the 1960s and 1970s, the similar-structure provided the framework for Zigler's expectation that persons with familial intellectual disability, as a group, would show the same level of performance on virtually every aspect of cognitive functioning when compared with typically developing persons at a similar general level of functioning (i.e., MA) (Mundy & Kasari, 1990; Weiss et al., 1986; Weisz, 1990; Weisz & Yeates, 1981; Zigler & Hodapp, 1986).

In a systematic and comprehensive review of empirical comparisons between the performance of persons with intellectual disability and that of typically developing children on Piagetian-type tasks, Weisz and Yeates (1981) found that the similar-structure hypothesis was supported in 90% of the 39 comparisons in the literature in which the appropriate developmental methodological strategies of matching the groups by MA and excluding persons with any indication of organic impairments or genetic anomalies were utilized. Thus, consistent with Zigler's hypothesis that familial intellectual disability is simply a slowed version of typical development, the findings across these studies indicated similar levels of performance by this group on Piagetian tasks in relation to typically developmentally children of the same level of functioning (i.e., MA rather than CA). However, the veracity of the similar-structure hypothesis was questioned by Weiss

et al. (1986) in a subsequent analysis of evidence from studies of information-processing, as persons with familial intellectual disability displayed inferior performance as compared with appropriately MA-matched typically developing persons on some tasks of memory, discrimination learning, and learning set, but not on other aspects of information-processing such as concept usage and incidental learning.

Weiss et al.'s (1986) challenge to the similar-structure hypothesis highlights the need to consider the implications of findings of deficits in cognitive functioning among persons with familial intellectual disability as compared with typically developing persons, an occurrence that appears to be more likely with the more precise methodologies associated with study of information-processing (Mundy & Kasari, 1990). Both the areas of functioning and the specific tasks that are associated with the evidence of impairment may be informative about the specific source of the problems on the specific test and about broader issues regarding the general performance of persons with intellectual disability. For example, the number of domains in which differences were found were limited and may be due to extenuating, and potentially confounding, factors in the relevant paradigms as impaired performance only appeared to be found in situations in which the tasks were long and repetitive, lacked ecological validity, and occurred in situations in which certain motivational and personality characteristics can affect performance. This is consistent with Zigler's early conceptualization of the various ways that the effects of a lifetime of experiences of failure affects the performance on experimental and real-life tasks by persons with intellectual disability. In extending this notion of the influence of personality and motivational factors on performance to highlight a "helpless" style of behavior that arises from a lifetime of failure, Weisz (1990) suggested that the discrepancies in performance in the studies cited by Weiss et al. may have been a function of the approach to the task at hand, rather than inherent differences in cognitive processing. Thus, the finding of these group differences seem more of an opportunity to further fine-tune Zigler's point of the necessity of understanding the whole person with intellectual disability than a contradiction of Zigler's similar-structure hypothesis as applied to persons with familial intellectual disability.

Cicchetti's Expansion of the Developmental Approach to Persons With Organic Etiologies: A Focus on Persons With Down Syndrome

Barely a decade after Zigler's (1967) original articulation of the developmental approach to intellectual disability,

Dante Cicchetti and colleagues (Cicchetti & Serafica, 1981; Cicchetti & Sroufe, 1976, 1978) provided its most compelling and far-reaching extension by initiating a program of research with children with Down syndrome that was based in developmental theory and principles. Although their work was focused exclusively on persons with Down syndrome, Cicchetti and colleagues (Cicchetti & Ganiban, 1990; Cicchetti & Pogge-Hesse, 1982) raised the possibility that these principles and theories could be applied to all persons with an organic etiology rather than just to persons with familial intellectual disability as argued by Zigler. As was the case with Zigler's seminal work, Cicchetti and colleagues' innovations were largely based in Heinz Werner's (1957) organismic-developmental approach with its overarching and universal organizational principles that provide meaning to the patterns of development (for a discussion, see Cicchetti & Ganiban, 1990). However, their application of classic developmental theory to persons with Down syndrome and, thereby essentially, to all persons with organically based intellectual disability provided both a much expanded view of development and a more stringent test of the universality of developmental theory than did Zigler's iteration of the developmental approach.

Cicchetti and Pogge-Hesse (1982) designated Zigler's developmental approach to intellectual disability as a conservative one in that it only applied to persons with familial intellectual disability as he excluded those with organically based intellectual disability. The rationale for this exclusion was that "if the etiology of the phenotypic intelligence (as measured by an IQ) of two groups differs, it is far from logical to assert that the course of development is the same, or even that similar contents in the behaviors of two such differing individuals are mediated by exactly the same cognitive processes" (Zigler, 1969, p. 533). In contrast to Zigler's conservative application of developmental theory and principles solely to familial persons with intellectual disability and their apparent typical patterns of development, Cicchetti and colleagues' expanded theory was focused on the search for coherence even across patterns of development that did not conform either to the norm or to the classic developmental theories of Piaget, Werner, and others. This more liberal framework transformed the developmental study of intellectual disability with its de facto inclusiveness of essentially all persons with intellectual disability under the umbrella of universal developmental principles while maintaining the essential ingredient of differentiating among etiologies. The unique patterns of relative strengths and weaknesses across areas of functioning for each specific etiological

group could be examined in relation to underlying patterns of development. Rather than delimiting the developmental approach and dismissing the unique development of specific etiological groups as different or impaired, Cicchetti and colleagues (Cicchetti & Ganiban, 1990; Cicchetti & Pogge-Hesse, 1982) sought coherence even as the developmental patterns diverged considerably from the typical (also, see Hodapp, Burack, & Zigler, 1990; Hodapp & Zigler, 1990; Wagner, Ganiban, & Cicchetti, 1990). For example, Cicchetti and Pogge-Hesse argued poignantly that the deficit approach "overlooks the possibility that the behavior and development of retarded children is organized, adaptive, and integrated—just as is the case for nonretarded children and infants. We know that they are retarded; the important and challenging research questions concern the developmental process" (p. 279). They concluded that "organically retarded persons are not only 'different' from nonretarded persons, they are organized in their own right . . ." (p. 313).

The centrality of this framework to the emergence of the developmental approach to the study of intellectual disability was highlighted by the inclusion of theoretically based review chapters by Cicchetti and colleagues (Cicchetti & Ganiban, 1990; Cicchetti & Pogge-Hesse, 1982) in the two initial edited volumes by Zigler and colleagues on the developmental approach (Hodapp, Burack, and Zigler, 1990; Zigler & Balla, 1982) and in a volume edited by Cicchetti and Beeghly (1990) devoted to the development of children with Down syndrome. Concordantly, Hodapp and Zigler's (1990) reframing of the conservative developmental approach to fit with the evidence from persons with Down syndrome (and, by extension, persons with other organic etiologies) in the opening chapter of the Cicchetti and Beeghly volume highlighted that the task of the expanded developmental study of intellectual disability was to seek developmental patterns across all etiological groups.

In broadening the cache of persons with intellectual disability to be studied within the context of the developmental approach, this more liberal developmental approach served to extend the relevance of classic developmental theories by implicitly challenging the common critiques of their deficiencies in accounting for group or individual differences (see Bronfenbrenner et al., 1986; Kessen, 1984). The study of Down syndrome and other etiological groups, with their own specific patterns of development, provides unique opportunities, or experiments of nature, to ask any number of essential questions such as which developmental patterns are necessary and which are happenstance? What skills are related to each

other throughout development and how closely need they be related? Where are alternative developmental pathways possible? Thus, Cicchetti and Pogge-Hesse (1982) argue that the orientation of the discussion about abilities needs to be changed so that “we should study not “does x have y” but rather “in what way does x use y” and does a particular mental structure interact with other mental structures in the same way in one . . . group as in another? (p. 313)”

One approach to this type of more complex, or liberal developmental, framework among persons with intellectual disability was initially suggested by Mundy, Seibert, and Hogan (1984) who borrowed the notion of local homologies, or developmental relationships across tasks that require underlying capacities, from the work on relationships across sensorimotor abilities among typically developing infants by Bates, Benigni, Bretherton, Camaioni, and Volterra (1979). Hodapp and colleagues (Hodapp, Burack, & Zigler, 1990; Hodapp & Zigler, 1990) proposed that this notion of local homologies may be essential to reconciling within an organizational developmental framework the apparent discrepancies, or lack of similar-structure (Weisz & Yeates, 1981), across areas of functioning that are found among persons with Down syndrome and other organic etiologies. Thus, this framework imposes some semblance of conceptual coherence across areas of functioning amid the apparent disconnectedness associated with the myriad of profiles of relative areas of strengths and weakness associated with the different etiologies.

In considering the various contributors to the complexity of the developmental approach to the study of persons with Down syndrome and other organic etiologies, Cicchetti and colleagues (Cicchetti & Ganiban, 1982; Cicchetti & Pogge-Hesse, 1982) highlighted that etiological groups could provide specific insights into the ways that both genes and organic function lead to behavioral outcomes. This notion is foundational to the contemporary notion of genes-brain-behaviors with specific etiological groups, each of which provides the opportunity for understanding the roles of specific genes and genetic structure as well as of organic function on behavior. As predicted by Cicchetti and colleagues, these endeavors would be enhanced by advances in technology and methodology that both allow for more extensive and precise groupings of persons with organic etiology and more sophisticated science with increasingly fine-tuned behavioral, physiological, and neuropsychological paradigms.

One particularly compelling expression of Cicchetti’s vision was the “neuroconstructivist” approach that was promoted by Annette Karmiloff-Smith (1998, 2009; Mareschal et al., 2007; Westermann et al., 2007). Building

upon the Piagetian notion of constructivism, with the child as an active agent in constructing his or her own cognitive structures through exploration and engagement with the environment, the neuroconstructivist model was developed and applied to children with organic etiologies associated with intellectual disability. Coined by Karmiloff-Smith (1998), neuroconstructivism brought together Piagetian constructivism with the emerging research on experience-driven neural plasticity. Karmiloff-Smith argued that to understand the cognitive profiles of organic etiologies, researchers needed to consider them within the context of the cascade of neural development. Using the example of Williams syndrome, Karmiloff-Smith et al. (1997) argued that rather than framing this syndrome as a modular disorder with spared language and impaired visual-spatial modules, the unique cognitive profile associated with it is the product of a unique developmental trajectory. As a corollary, Cornish and colleagues (Cornish, Cole, Longhi, Karmiloff-Smith, & Scerif, 2013; Cornish, Steele, Monteiro, Karmiloff-Smith, & Scerif, 2012; for a review, see Cornish & Wilding, 2010) highlighted that groups of persons with different organic etiologies might exhibit different attentional profiles and inhibitory control beginning in infancy, which in turn contribute to different developmental pathways. The neuroconstructivist framework then casts each of the organic etiologies as having genetically determined constraints, which manifest as subtle differences in preferences and biases early in development, steering the child down an etiology-specific developmental pathway that is nonetheless constructed by the child (e.g., for discussions regarding fragile X, see Cornish, Bertone, Kogan, & Scerif, 2012; Cornish, Scerif, & Karmiloff-Smith, 2007; for a discussion regarding Williams syndrome, see Elsabbagh & Karmiloff-Smith, 2012).

In promoting the neuroconstructivist approach, Karmiloff-Smith and colleagues challenge the virtually exclusive dependence on cross-sectional methodologies, with their snapshots of performance at a specific moment in the lifespan, in the quest to better understand intact or impaired performance in the study of specific etiologies. This is consistent with the notion that performance is always relative—relative not only to another group but most important also to past and future performance. Ideally, developmental disorders (and development in general) should be examined longitudinally, although as Karmiloff-Smith (2009) acknowledges the measurement of change over time often needs to be simulated using regression-based approaches due to the pragmatic difficulties inherent in longitudinal studies. The extent to which our understanding of development is constrained by cross-sectional methodology is highlighted

by Cornish et al. (2013) who depicted both a trajectory mapping correlational study and a one-year longitudinal follow-up study of attentional control and working memory among children with fragile X. Whereas the data from the cross-sectional trajectory mapping suggested developmental arrest in working memory and a dramatically slower (relative to typical) but positive slope in relation to MA for attentional control, the longitudinal data indicated similar slopes of improvement for both skills in relation to the performance of the typically developing children.

Another example of the contemporary realization of the need for increasingly sophisticated methodology and precise understanding of the development of gene–brain–behavior relationship within a specific etiological group is Barbara Landau’s (2012; Landau & Hoffman, 2007) work on the ways that developmental time is essential to the organization and development of spatial representations among persons with Williams syndrome as well as among typically developing persons. Landau argues that one of the ways in which, genetic etiologies contribute to intellectual disability is in the disruption of the rate of development, but emphasizes that the mapping of developmental timetables in these etiologies needs to be considered in relation to typical development. To illustrate this, Landau argues that the spatial capacities that are strong among persons with Williams syndrome are associated with the ventral stream, which emerges relatively early in the typically developing child, whereas the capacities that are weaker are associated with the dorsal stream, which develops later in typical development. For example, children with Williams syndrome (MA 4–6 years) perform at or above MA expectations on tasks used to measure aspects of spatial abilities that involve the ventral stream, which develops early in typical development (e.g., perceptual matching). Conversely, differences in performance emerge on dorsal stream tasks for which development is protracted in typical development. Based on this evidence, Landau hypothesized that the developmental trajectory of the dorsal stream is slower than that of the ventral stream for persons with Williams syndrome and that it is the exaggerated asynchronous development of the two streams that contributes to their unique profile of spatial abilities.

THE IMPORTANCE OF MENTAL AGE

Considering Developmental Level

In Zigler’s (1967, 1969) critique of the prevailing defect approach of the time, the most obvious, and thereby the most troubling, of the essential methodological flaws that

he cited as inherent to the defect theorists’ research and findings was the emphasis on CA as the point of comparison between persons with and without intellectual disability. The defect theories were based largely on evidence from studies in which the performance of a group of persons with intellectual disability was compared with that of a group of typically developing persons who were matched, or deemed similar, to the first group on the basis of CA. Since persons with intellectual disability are, by definition, lower functioning than those without intellectual disability of the same CA, the outcome of such research is obvious. The persons with intellectual disability would always be expected to perform worse on any experimental task that is appropriate for the level of functioning of the participants and that is sufficiently sensitive to differentiate between groups of persons within the relevant range of level of functioning. Yet, despite their inevitability, findings of group differences based in this methodology were considered essential evidence by the defect theorists in their promulgation of the various impairments that were hypothesized as the core deficits of intellectual disability.

In highlighting one example of the extent to which CA matching research was integral to promoting the defect approach, Iarocci, Burack, and colleagues (Burack et al., 2001; Burack, Dawkins, et al., 2012; Iarocci & Burack, 1998; Iarocci et al., 2012) demonstrated that the commonly accepted notion of a core defect in attentional functioning, which was perpetuated from the 1960s even well into the 1990s, was based largely on empirical evidence in which matching was exclusively based on CA. In addressing this claim in a review of the rather limited empirical evidence available in which the group of persons with intellectual disability were homogeneous with regard to familial status and were matched to the comparison group of typically developing children on the appropriate basis of MA, Iarocci and Burack found no evidence for any attention deficit. Rather, they concluded that the defect approach researchers of attention had promoted “fatally” flawed research and had simply and inevitably found that lower functioning persons functioned at lower levels than higher functioning persons.

Iarocci and Burack (1998) acknowledged that the failure to find impairments in attentional functioning among persons with familial intellectual disability certainly does not rule out the possibility of finding deficits either among persons with familial intellectual disability on different components of attention, with different tasks, or at different ages; or among groups of persons with specific organic etiologies, some of which have been linked to preliminary developmental patterns of attention problems (for recent reviews, see Cornish & Wilding, 2010; Iarocci et al., 2012).

Yet, they and their colleagues highlight that the use of essentially problematic methodology of CA-matching led to the perpetuation of a myth of attention deficit in the study of intellectual disability (Burack et al., 2001; Burack, Dawkins, et al., 2012).

Given the obvious problems with CA-matching, what is the possible justification for the decades-long perpetuation for this fatally flawed methodology? One common argument for CA matching is that it allows for the identification of “spared abilities” in specific areas of functioning (for a discussion of the arguments in favor of CA matching and their fatal flaws, see Burack, Russo, et al., 2012). The proponents of CA matching argue that if persons with intellectual disability perform similarly to a CA-matched group of typically developing persons in a specific area of functioning then functioning in that area could be considered to be uniquely spared. Unfortunately, this reasoning is inconsistent with the basics of experimental developmental research. The failure to find differences between groups can rarely, if ever, be considered unqualified, or even strong, evidence that the groups perform similarly. Rather, this type of finding is more likely the consequence of one or more methodological constraints. In the case of research with persons with intellectual disability, the failure to find group differences is often because the participants do not function at the developmental levels at which differences in the specific area of functioning might optimally be found. For example, if an area of functioning emerges at age X, testing participants considerably prior to age X would not elicit group (or individual) differences since the area of functioning would not have yet developed for even the typically developing persons, and therefore both the typically developing children and children with intellectual disability would display apparently low levels of performance. Concordantly, differences would also be unlikely to be elicited with participants who are significantly older than X as the persons with intellectual disability would have had ample opportunity to attain the requisite skills even if their development in the specific area of functioning is slower as compared with the typically developing persons. Even in the case when the participants are all at the ideal age to study the emergent area of functioning, differences might not be found because of 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 sparing of abilities is more likely a consequent of problematic methodology than of any meaningful characteristic of the specific group (Burack, Russo, et al., 2012).

The most obvious matching strategy for comparing groups with different a priori levels of functioning, such as indicated by IQ, is to find a way to equate the groups on a general criterion that is relevant to the aspect of functioning that is being studied. In the study of cognitive and cognitive-related abilities among persons with intellectual disability as compared with those without intellectual disability, the relevant criterion is level of cognitive functioning, or MA, at the time of testing. In this context, the task of the researcher is to find groups of persons with and without intellectual disability that are similar on these measures, which can be accomplished only with groups that are considerably discrepant with regard to CA. Although the persons with intellectual disability are inevitably chronologically older, the strategy of matching by MA diminishes, or even eliminates, the potential criticism that findings of deficits among the persons with intellectual disability can 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 intellectual disability is evident even when general functioning is equated. This is, of course, a much stronger argument than when the a priori level of functioning differs between the groups, as in the case of CA matching. Thus, the original call to match by MA (see Zigler, 1967, 1969; Zigler & Balla, 1982; Zigler & Hodapp, 1986), or general developmental level, was a meaningful improvement over the CA-matching strategies of yesteryear since the apparent deficits of persons with intellectual disability could not be simply attributed to a priori levels of development and functioning.

Although matching on the basis of MA represents an advance in the field, the process of matching is inherently flawed (Burack, Russo, et al., 2012; Jarrold & Brock, 2004) as the task of equating such different groups of persons is plagued by the fact that the groups are, in fact, essentially different. In highlighting this point, Burack, Russo et al. note that even in the imaginary scenario in which a person with intellectual disability and a typically developing child would have the exact same MA for a moment in time, they would differ from each other in so many different ways. By definition, the typically developing child would have attained the shared MA at a faster rate and at a younger age, whereas the person with intellectual disability would have lived longer and be more physically and physiologically mature. In addition to the age differences, the two would have had very different life experiences, especially with regard to support and encouragement, successes and failures, and the nature of familial and other relationships. Their futures would also diverge.

In the hypothetical case of an ideal test with perfect sensitivity and reliability in which the two children were perfectly commensurate with regard to level of functioning at a point in time, the typically developing child would surge ahead the moment after.

The inherent limitations in MA matching in the study of persons with intellectual disability are particularly exposed in the study of specific etiological groups, as the unique profiles observed for each of them relegate general scores of MA as insufficient to mitigate against claims of a priori differences between the groups (e.g., Karmiloff-Smith, 2009). In a relevant paper on matching in research with persons with autism, Burack et al. (2004) point out that “no choice of comparison group or matching strategy is perfect, but rather needs to be determined by specific research objectives and theoretical questions” (p. 65). In this context, they and others (e.g., for a detailed discussion, see Mervis & Klein-Tasman, 2004) highlight the need to utilize matching measures that are even more precise than general 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 in the area of functioning related to the experimental task for one of the groups. As an almost universal manifestation of this approach, researchers typically used measures of performance or nonverbal measures to match groups for studies that involve visuo-spatial processing, whereas verbal measures are most often used as the matching criterion for studies in which verbal abilities are central. However, this more precise strategy is still replete with problems as many experimental tasks involve both visual-spatial and verbal reasoning. Furthermore, the matching measures often lack precision, are often either particularly broad indicators of functioning, or are only tangentially related to the experimental task so that they are only minimally more relevant than general MA to the skills for which they are supposed to control. Despite the seminal methodological advances that arose from Zigler’s articulation of MA, rather than CA, as the essential criterion for matching groups in studies with persons with intellectual disability and the subsequent imposition of increasingly precise indicators of matching, Burack, Russo, et al. (2012) suggest that the discussion should be less about “the way” or even “best way” to match, but rather, more humbly, the “least bad way” for the specific context of a given study. This acknowledgment of the inherent limitations with matching strategies is highlighted by the calls by

some developmentally-oriented researchers to supplement, or even supplant, research methodologies based on the matching of groups with those involving regression or longitudinal paradigms (Cornish et al., 2013; Jarrold & Brock, 2004; Karmiloff-Smith, 2009).

THE STUDY OF THE “WHOLE PERSON” WITH INTELLECTUAL DISABILITY

The fourth of Zigler’s (1967, 1969) primary contributions to the development of the developmental position best highlights the humanistic aspect of Zigler’s developmental work (for a discussion, see Burack, 1997). Amid the field’s obsessive focus on identifying a central, and inevitably cognitive-based, defect to explain intellectual disability, Zigler further distanced himself from his contemporaries by highlighting the need to focus on the persons with intellectual disability rather than on the phenomenon of intellectual disability and on all the domains of functioning that comprise the whole person rather than on just a specific area (or areas) of deficit. He argued that being a person with intellectual disability inevitably involved life experiences that would affect the way the person behaves and performs in all aspects of their life. However, true to his developmental outlook, Zigler even framed this apparently population-specific consideration within the context of universal developmental issues. The focus wasn’t exclusively on the effects of intellectual disability or even on the life experiences of being a person with intellectual disability, but rather was framed within the context of a lifetime of failure, lowered expectations, and, in keeping with the times, institutionalization, all of which were relevant, but not exclusive, issues in the lives of persons with intellectual disability. As with other groups who shared some of these same life history backgrounds, such as persons from historically oppressed and particularly low SES groups, Zigler imagined that certain personality characteristics related to the motivation of the individuals with intellectual disability would develop in relation to the life circumstances and would permeate every aspect of the person’s functioning. Although Zigler cited the personality-motivational characteristics as explanations for lower than expected performance for persons with familial intellectual disability on experimental tasks and how they could interfere with optimal task performance, these traits are as, if not more, relevant to virtually every aspect of any person’s daily life.

These hypothesized interrelated traits include overdependency, low self-esteem, wariness of others, limited

mastery motivation, reinforcement-seeking, and outer-directedness (rather than innerdirectedness) (for reviews, see Merighi, Edison, & Zigler, 1990; Zigler & Burack, 1989; Zigler & Hodapp, 1986). Each of these styles of behaviors can be seen as a natural outcome for someone one who has lived with constant and consistent failure, little if any positive reinforcement, considerable criticism, lowered expectations, and other inevitable experiences of someone who functions outside the realm of society or fails to meet societal expectations. Yet they are also part of a more universal humanistic approach to understanding behavior as the same traits found among persons with intellectual disability or even others on the fringe are also found among persons in the mainstream society who have experienced failure. From a developmental perspective, the essential point of these characteristics is that they can even be adaptive in the appropriate context. For example, almost all of us utilize an outerdirected strategy in checking what others do when placed in situations that are particularly novel, dining in a restaurant of a different culture for the first time, or complex, dining at a particularly elegant event in which a multitude of dishes, glasses and utensils are provided. In these situations, we typically look around to see what is expected and act in relation to how others do in order not to embarrass ourselves, whereas in other more comfortable or common situations, we tend to behave in a more innerdirected way. In contrast, Zigler noted that persons with intellectual disability rely on the outerdirected approach even in more common situations in which they could perform well or at least accomplish what is expected without looking to others for cues as to how to behave. Bybee and Zigler (1992) highlighted this point with their finding that typically developing children used an outerdirected strategy in which they copied and otherwise looked to the experimenter's production of a picture using stickers for a novel or difficult task demand which they could not have otherwise fulfilled, whereas children with intellectual disability of the same MA used an outerdirected approach even on a simpler task that they should have been able to perform without external help (for a relevant review, see Bybee & Zigler, 1998).

The extent to which personality-motivational factors influence the lives of persons with intellectual disability, especially those with familial intellectual disability, is highlighted by findings that they are the best predictors of success and ability to integrate into mainstream aspects of society among persons within the range of mild intellectual disability (for reviews, see MacLean, 1997; Zigler & Hodapp, 1986). Consistent with the notion that mild intellectual impairment associated with familial

intellectual disability simply reflects persons at the lower end of the natural continuum of the bell curve of intelligence, many persons in this grouping are able to hold jobs, live independently, and even start their own families (Bernheimer, Keogh, & Guthrie, 2006; Keogh et al., 2004; West, Wehman, & Wehman, 2005). As is the case with other persons, success is contingent primarily on extracognitive factors such as those related to personality and motivation (Merighi et al., 1990; Seltzer et al., 2009).

Although Zigler's (1967, 1969) initial broadening of the field of intellectual disability in the 1960s and 1970s to include extracognitive factors was focused almost solely on personality and motivational factors, it can be seen as the beginning of a more humanistic approach to research and scholarly understanding the whole person with intellectual disability. With the emphasis on the whole person, issues regarding the social and emotional lives of persons with intellectual disability became a priority for researchers, a timely occurrence in light of the relatively contemporaneous (and soon-to-follow) revelations of the atrocious conditions in which many persons with intellectual disability lived (e.g., in 1972, Geraldo Rivera exposed the conditions of the Willowbrook State School, a state-funded institution on Staten Island, New York, that housed 6,000 persons with intellectual disability) and the emergent emphasis on social adaptation in the essential (*DSM* and *AAMD*) diagnostic classification systems.

As with Zigler's other contributions to the developmental approach, the emphasis on personality and motivational factors and the whole person was expanded in many ways. The most notable is that Zigler's (1967, 1969) specific focus on these issues with regard to individuals with familial intellectual disability have since been applied to those with specific organic etiologies. Again, Cicchetti and colleagues provided early extensions of Zigler's dictum, as they focused on the temperament and socioemotional development specifically among children with Down syndrome (for extensive reviews, see Cicchetti & Beeghly, 1990; Cicchetti & Ganiban, 1990; Cicchetti & Sroufe, 1978). This type of etiology-specific delineation is evidence of the advances in understanding the whole person in relation to essential issues that affect and are affected by development.

In this section, we highlight the advances and some of the difficulties in the specific study of three issues, social competence, language, and family relationships, that have been considered in relation to specific etiological groups and that reflect a commitment to understanding the social-emotional well-being of the whole person. Social competence represents the individual's abilities and

motivation to successfully interact in meaningful ways with others. Language reflects the case of a specific area of functioning that is essentially interrelated with social development. Family relationships involve issues of the reciprocal relationship between the presence of the child with intellectual disability and the family context. By disentangling these and related issues, we can better understand the transactions among the different systems that affect the whole person as well as between the individual and their immediate and broader systems (Bronfenbrenner, 1979). In the following sections, we consider the research on these components of the whole persons among individuals with the most commonly studied etiologies, Down syndrome, fragile X syndrome, Williams syndrome, and Prader-Willi syndrome.

Social Competence

Social competence is a key construct within the study of social development because it captures the dynamic relation between cognitive and social factors as they are applied to social adaptation (Iarocci, Yager, Rombough, & McGlaughlin, 2008). Although no operational definition of social competence is universally accepted, the term spans both socially adaptive behaviors as well as more basic social cognitive processes (Yager & Iarocci, 2013). It has been conceptualized as occurring at various levels of analysis—at the level of cognition (e.g., perspective-taking); overt behaviors/skills (e.g., use of eye contact); and outcome (e.g., social status, peer acceptance).

Developmental and contextual changes add further complexity to the study of social competence. It is both a developmental phenomenon that can be measured over the course of a child’s development (i.e., ontogenesis) as well as a characteristic of a particular social encounter for which the time scale is in the order of seconds/minutes (i.e., microgenesis). Thus, continuities and discontinuities in the development of social competence are expected; children are better able to coordinate abilities and take advantage of resources with increasing age but may be less competent at certain developmental stages or in specific social contexts. What constitutes socially competent behavior changes depending on the age of the child as well as the context within which the child is evaluated (e.g., with parent or peers).

The same set of social skills are not available to the infant as they are to the adolescent. Thus, age-appropriate assessment in which the measure’s focus is targeted to particular developmental periods are essential. For example, in infancy the quality of the parent–child interaction may

be a particularly sensitive index of social competence as the interrelations between infant cues and parent sensitivity to the infant’s cues improve the odds of survival as well as the social-emotional development of the infant. Peer interactions may be especially salient in middle childhood, whereas intimate relationships and community integration are paramount in adolescence.

In considering these aspects of social competence as well as other aspects of social development across populations, Yager and Iarocci (2013) note even persons with severe social impairment may be intensely aware of their social disconnectedness, experience distressing loneliness, and show a desire for greater social interaction. Accordingly, they emphasize that social motivation and social understanding may be relatively independent and measurable aspects of social competence. Within this framework, we use Rose-Krasnor’s (1997) “prism model” to guide our understanding of the literature on social competence among persons with intellectual disability as it provides a useful framework for conceptualizing social competence and its levels of analysis. According to the model, social competence at a theoretical level is broadly defined as overall effectiveness in meeting short- and long-term social developmental needs/goals. Thus, social competence is viewed as a higher-order, organizing construct with transactional, context-dependent, and goal-specific characteristics. It cannot be reduced to any single index or skill but requires the active, skillful coordination of multiple processes as well as contextual factors to adequately meet the social demands of a particular situation (Iarocci, Yager, & Elfers, 2007; Iarocci, Yager, Rombough, & Mclaughlin, 2008). However, identifying and measuring a representative sampling of abilities/behaviors at the social motivation and skills level are useful in order to “tap into” the broader theoretical construct of social competence.

According to the prism model, social competence can be studied empirically at two distinct levels of analysis: the *index* level and the *motivation/skills* level (Rose-Krasnor, 1997). The index level represents real-life summary indicators of social competence (e.g., attachment security, peer acceptance, or employment success) that are considered to be situation/context-specific (e.g., with peers vs. with family). Although generally indicative of social competence, these indexes do not provide detailed information about the social presentation of an individual. Their utility in assessment is primarily as outcome measures or gross screening tools (e.g., identifying individuals who may be socially impaired). The *motivation/skills* level is comprised of the underlying dispositions and abilities that provide the “building blocks” of social interactions (e.g., social

motivations and social skills). Social skills include both social-cognitive abilities (e.g., perception and processing of social stimuli) and the more overt, observable social behaviors (e.g., eye contact, conversation ability). Any given social motivation or skill may be relevant across contexts or prove particularly valuable within a specific social context. In general, social motivations and skills tend to be more accessible to assessment methods (as compared with the indexes) and are commonly measured with performance-based laboratory tasks (e.g., tests of emotion recognition) or observation-based rating scales.

In this brief review of research on social competence among specific etiological groups of persons with intellectual disability, we include studies that are focused on the index level and those that are focused on the motivation/skills level. At the index level, the studies provide information about the individual's social behavior for adaptation, whereas those at the motivation/skills level contribute to an understanding of the socio-cognitive processes that underpin social behaviors.

Social Competence Among Persons With Fragile X Syndrome

Children with fragile X are often diagnosed with ADHD as they show poor attention span, impulsivity, and high motor activity. With age, hyperactivity and impulsivity subside yet attention deficits and inhibitory control problems persist (Cornish et al., 2008). High rates of autism spectrum disorder are also noted in this group along with considerable overlap of symptoms, although those with fragile X and autism display significantly less impairment on measurements of social response on the Autism Diagnostic Observation Schedule (ADOS), including the areas of Response to Joint Attention, Responsive Social Smile, and Facial Expressions (Wolf et al., 2012). For example, children with fragile X tend to show gaze avoidance, hyperarousal, repetitive activity, and motor stereotypy. Expressive language deficits are also common and include perseverative speech, echolalia, self-talking, and cluttering (Fletcher et al., 2007). Children with fragile X are also shy and at higher risk of developing social anxiety (Teisl, Reiss, & Mazzocco, 1999).

At the Index level, individuals with fragile X show relatively good adaptive skills and many are able to engage in occupational activities (Fletcher et al., 2007). However, they continue to show problems with temper outbursts as well as obsessional and aggressive behaviors well into adulthood (Howlin & Udwin, 2002).

At the motivation/skills level, children with fragile X are reported to show less approach, persistence, and

adaptability in social interactions. Individuals with fragile X are described as socially anxious and withdrawn, and tend to avoid eye contact and have limited interaction with peers (Cornish et al. 2001; Kaufmann et al. 2004). In an analysis of scores on the Autism Diagnostic Observation Schedule (ADOS), Wolf et al. (2012) found that participants with both fragile X and autism were capable of appropriately responding to social bids made by others as indicated by their performance on the subtests of Responsive Social Smiling, Facial Expressions, Response to Joint Attention, but showed poor social initiative as indexed by the of Showing, Requesting, and Initiation of Joint Attention. Accordingly, Wolf et al. (2012) argued that the social difficulty in fragile X has more to do with a lack of prosocial initiative rather than any lack or impairment of social ability or social awareness. They speculated that the lack of initiative in making social bids was likely related to the high level of social anxiety in this group.

Initial evidence regarding the processing of emotions, faces, and other social information among persons with fragile X underscores the importance of considering developmental level, rather than CA, as patterns of performance which appear to reflect universal impairments or differences in relation to that of same-aged peers are considerably more nuanced in relation to MA-appropriate levels. For example, Shaw and Porter (2013) found that participants with fragile X (mean CA of 24.8 years and mean MA of 8.4 years) displayed specific emotion recognition deficits for angry and neutral, but not happy or fearful, faces as compared with both MA-matched (mean MA of 9.2 years) and CA-matched (mean CA of 24.5 years) typically developing persons, but developmentally appropriate visual scanning of emotional faces. Although the participants with fragile X displayed reduced attention to the eyes and different visual scan patterns of emotional faces as compared with the CA-matched typically developing individuals, their scanning patterns were commensurate with those of the MA-matched typically developing participants. The complexity in the relation between behavior and processes is highlighted by Hagan, Hoeft, Mackey, Mobbs, and Reiss's (2008) finding that 10 high-functioning females with the fragile X full mutation (mean CA of 16.4 years and IQ of 91) as compared with 10 typically developing girls matched on age but not IQ (mean CA of 15.2 and mean IQ of 106) showed intact emotion recognition despite a potentially disrupted neural circuit that modulates emotional responses to faces even when differences in IQ and accuracy of responses were statistically controlled. In a related study of visual scan paths, Williams, Porter, and Langdon (2013) found that

participants with fragile X (mean CA of 23 years and MA of 8.7 years) generally attended to social information overall to a similar degree as MA- (mean MA of 8.7 years) and CA- (mean CA of 23.2 years) matched typically developing participants, but were faster to disengage their attention away from social information in naturalistic static social scenes as compared with these groups as well as with a group of participants with Williams syndrome. Williams et al. concluded that the faster disengagement of attention away from social information was evidence of the avoidance of social stimuli among persons with fragile X.

In a study of higher order skills relevant to social understanding, Losh et al. (2012) examined theory of mind and pragmatic language skills among boys with fragile X-only, boys with fragile X and autism, boys with autism only, boys with Down syndrome, and typically developing boys. They found that both the participants with autism-only and the participants with both fragile X and autism performed most poorly, whereas the boys with fragile X-only did not differ significantly from the group of boys with Down syndrome or the MA-matched typically developing boys. Moreover, for all the groups, better theory of mind scores were associated with more pragmatic language competence as indexed by the Pragmatic Judgment subtest of the Comprehensive Assessment of Spoken Language. Losh et al.'s conclusion that the boys with fragile X-only showed relative strengths in theory of mind and pragmatic language and therefore that the deficits in these areas may be specific to those with both fragile X and autism highlights the social differences between the two subgroups of fragile X and suggests that any social problems associated with fragile X per se are not likely associated with higher-order cognition.

In summary of the work on social competence among persons with fragile X, the few preliminary studies on index level suggest good adaptive and occupational skills, but also behavioral problems that may interfere with overall social adjustment. At the motivational/skills level, social avoidance and anxiety appear prominent, yet unusual social attention processing is also present. Preliminary, but complex, profiles of both age-appropriate and impaired emotion recognition and theory of mind emerged, depending on what was being measured and with what task. Difficulties with theory of mind and pragmatic language tasks were specifically associated with fragile X, but only when co-occurring with autism.

Social Competence Among Persons With Down Syndrome

Children with Down syndrome typically fare better than children with other etiologies with regard to certain

aspects of social competence. For example, at the index level they show generally good social adaptation and positive social outcomes (Rosner, Hodapp, Fidler, Sagan, & Dykens, 2004), whereas the profile is mixed at the motivation/skills level. Rosner et al. (2004) studied 177 persons with intellectual disability aged 4–49 years and found that the number and quality of social activities and relationships as measured by the Child Behavior Check List (CBCL; Achenbach, 1991) was significantly better among participants with Down syndrome compared with groups of children with Prader-Willi and Williams syndrome, after both age and IQ were covaried in all between-group analyses to account for the fact that the participants with Down syndrome were approximately 4–5 years younger and 12–16 points lower in IQ than the other two groups. In particular, the children and youth with Down syndrome were reported to behave well with others, show involvement in organizations, and demonstrate good job skills. In apparent contrast, Guralnick, Connor, and Johnson (2009) focused on more fine-grained analyses of peer social contacts, networks, quality of play, degree of control of play, and the characteristics of playmates, and found fewer well-developed peer networks in comparison to a MA-matched group of typically developing children. Specific areas of difficulties noted were the children's level of involvement in play with peers, linkages to other settings (e.g., school), and control of play. The seeming inconsistencies between the studies simply highlight two essential methodological points—the type of measure (global vs. fine grained) and the comparison group (typically developing vs. developmentally disabled) need to be considered when interpreting findings.

With regard to the motivation/skills level, despite evidence of relatively good motivation and interest (e.g., face gazing and smiling behaviors) (Moore, Oates, Hobson, & Goodwin, 2002), difficulties have been noted in aspects of social cognition such as joint attention and theory of mind as compared with both MA matched typically developing children and children with nonspecific intellectual disabilities (for a review, see Iarocci et al., 2008). Children with Down syndrome were found to show fewer social referencing looks to their mothers in ambiguous situations (Kasari, Freeman, Mundy, & Sigman, 1995b) and make fewer attempts to direct their mother's attention across social situations (Fischer, 1987; Landry, Garner, Pirie, & Swank, 1994). Concordantly, Fidler (2005) found that toddlers with Down syndrome showed fewer nonverbal, instrumental requests than did typically developing and developmentally disabled toddlers matched on MA

and that these requests were related to the level of their problem-solving abilities.

Children with Down syndrome appear to show poorer emotion recognition relative to their cognitive abilities (Kasari, Freeman & Hughes, 2001; Kasari, Freeman & Bass, 2003), and appear to have particular difficulties with identifying and labeling fear and anger (Kasari & Freeman, 2001). Even when language demands were minimized with the use of nonverbal photo matching of expressions, Williams et al. (2005) found that children with Down syndrome had difficulty with fearful expressions. Concordantly, Wishart et al. (2007) found that persons with Down syndrome were particularly impaired on matching fearful expressions as compared with individuals with fragile X syndrome, those with nonspecific intellectual disability, and typically developing children. In addition to the difficulties recognizing emotions in others, children with Down syndrome have been found to show decreased emotional expressiveness. Early in infancy, they appear to have dampened smiles, shorter and less frequent displays of emotion, and longer delays to express emotion as compared with typically developing children (Emde & Brown, 1978; Thompson et al., 1985). Jahromi, Gulsrud, and Kasari (2008) found that children with Down syndrome generally showed more negative affect and difficulty regulating their emotions during challenging tasks as compared with typically developing children. The children with Down syndrome seemed to have a limited repertoire of behaviors to cope with frustration and, in fact, displayed more frustration and fewer cognitive self-soothing techniques. Consistent with the notion that personality-motivational characteristics can interfere with optimal performance among persons with intellectual disability, the children with Down syndrome oriented more to the experimenter without asking for help.

Despite the apparent difficulties in emotion recognition, expressivity and regulation, children with Down syndrome generally show more responsiveness to others (e.g., comforting) during distressing situations as compared with typically developing children. Although Kasari et al. (2003) found a poorer understanding of the concept of empathy among persons with Down syndrome, they reported an awareness of situations that are potentially distressing to others and the commission of appropriate actions to comfort others.

In adolescence and early adulthood, individuals with Down syndrome appear to become more withdrawn and show less pleasure or animation (Fletcher et al., 2007). The social withdrawal may be related to their higher risk of mental health problems such as the higher incidence

of depressive disorder, anxiety, and obsessive compulsive disorder among the young adults (Dykens, Shah, Sagun, Beck, & King, 2002; Fletcher et al., 2007). Early onset Alzheimer's disease is also more likely in this group and possibly has adverse effects on social functioning (Prasher, Chung, Hague, 1998).

In summary, at the index level, children and youth with Down syndrome appear to show relative strengths in social competence as compared with other developmentally delayed youths. However, the type of measure and the comparison group need to be considered when interpreting as these findings of strengths may not be found in comparison to typically developing children. At the motivation/skills level, social motivation, interest, and responsiveness appear to be relative strengths, whereas social-cognitive abilities and emotion recognition, expressivity and regulation may be areas of relative weakness.

Social Competence Among Persons With Williams Syndrome

Children with Williams syndrome exhibit "hyper-sociality" and tend to be overly friendly, engaging, and responsive to others (Fidler, Hepburn, Most, Philofsky, & Rogers, 2007; Mervis et al., 2003). The social approach behavior appears to be compulsive since these children's approaches to strangers are typically indiscriminant even when the children have knowledge of the approachability of the other (Frigerio et al., 2006), and most report being scared of strangers (Dykens, 2003). Children with Williams syndrome show intense and prolonged attention to faces to the detriment of attention to other aspects of their environment (Mervis et al., 2003), yet their emotion recognition skills are delayed (Fidler et al., 2007; Gagliardi et al., 2003).

At the index level, individuals with Williams syndrome appear to show relative weaknesses in adaptive daily living skills (Hahn, Fidler & Hepburn, 2014; Mervis et al. 2001; Plissart et al. 1994), although they improve into adulthood (Howlin, Elison, Udwin, & Stinton, 2010). Howlin et al. used both cross-sectional and longitudinal designs and found no evidence of cognitive or linguistic deterioration overtime that could account for poor adaptive functioning among persons with Williams syndrome. Rather, adaptive functioning (as measured by the Vineland Adaptive Behavior Scales) generally improved until age 50 years. Specifically, Communication domain scores were consistently lower than scores in other domains and the relations between Socialization and Daily Living scores varied with age. However, parent reports suggest that these

individuals are dependent on family members and struggle with physical and mental health problems in adulthood. For example, 239 parents of adults (age 18–56 years) with Williams syndrome surveyed by Howlin and Udwin (2006) reported high rates of physical health problems as well as increases in rates of mental health problems (depression or anxiety) with age. Educational and employment attainments were generally low and self-help skills were relatively poor. Most (62%) of the adults with Williams syndrome were still living at home, some with quite elderly parents.

In Rosner et al.’s (2004) comparison of participants with Down syndrome, Williams syndrome, and Prader-Willi syndrome, those with Williams syndrome scored lower on social competency on the CBCL as compared with participants with Down syndrome, but higher as compared with participants with Prader-Willi syndrome. Most persons with Williams syndrome experienced difficulty with social interaction and were socially isolated (Davies et al., 1998). In one early study, 96% of all the individuals were described as experiencing difficulty in making friends and 76% were reported as having few to no friends (Udwin, 1990). With regard to occupational functioning, those with Williams syndrome scored lowest on job skills using the CBCL, which includes both household chores and jobs outside the home (Rosner et al., 2004). Problems with distractibility, social disinhibition and overfriendliness, as well as anxiety have also been noted (Davies et al., 1997; Udwin et al., 1998).

At the motivation/skills level, children with Williams syndrome have been found to show unusually intense social interest and processing of faces and eyes. For example, children with Williams syndrome fixate on faces longer than verbal IQ matched typically developing children and are slower to disengage their gaze once fixated on eyes (Porter et al., 2010) or a face (Riby et al., 2010). At an even more general level, Williams, Porter, and Langdon (2013) also reported difficulties disengaging attention rather than attentional capture among individuals with Williams syndrome, after they manipulated the location of socially salient information within visual scenes. These studies suggest that some aspect of selective attention may be impaired and implicated in the social perception behavior of children with Williams syndrome.

Despite the high social interest and intense attention to faces, individuals with Williams syndrome display significant impairments in recognizing emotions (Lacroix, Guidetti, Roge, & Reilly, 2009). Although they look longer at social stimuli they do not appear to process all relevant aspects but, rather, show a bias toward positive elements. In a study on autonomic responsiveness to emotionally laden

images with social or nonsocial content, Plesa-Skwerer et al. (2011) found that the participants with Williams syndrome looked significantly longer at the social images as compared with images without social content and displayed reduced arousal to the negative social images as compared with the nonverbal MA-matched typically developing groups. Ironically, the awareness of emotional distress in others and heightened level of sensitivity to the emotional reactions of others (as evidenced by their high level of mimicry and imitation) does not appear to be applied to social decision making (Fidler et al., 2007).

Cognitive and language deficits seem to play a role in the social behavior of individuals with Williams syndrome. They appear to demonstrate difficulties in higher order social-cognitive functions such as atypical nonverbal communication, imagination, and problems in understanding the mental states of others (i.e., theory of mind; Plesa-Skwerer, Faja, Schofield, Verbalis, & Tager-Flusberg, 2006; Tager-Flusberg & Sullivan, 2000). Generally, people with Williams syndrome show about the same level of performance on various theory of mind tasks as other children with developmental disabilities (Plesa-Skwerer & Tager-Flusberg, 2006). Tager-Flusberg and Sullivan suggested that theory of mind functioning among persons with Williams syndrome is characterized by relative sparing in the social-perceptual component, the ability to make an immediate judgment of a person’s mental state, but impairment on the social-cognitive component. The social-pragmatic aspects of language (e.g., sensitivity of the communicative partner’s perspective, nonverbal and verbal cues, and conversational reciprocity, interpreting jokes) are poor and similar to those of persons with autism (Klein-Tasman, Mervis, Lord, & Phillips, 2007; Stojanovik, 2006; Stojanovik & James, 2006).

The social approach behaviors of people with Williams syndrome may be due to a reduced ability to inhibit the urge to socially interact with others, suggesting a general rather than a social deficit in inhibition (Porter et al., 2007). For example, in examining the variability in the social approach behaviors of children with Williams syndrome ranging from 6 to 15 years of age with a cluster analysis, Little et al. (2013) identified social approach behavior subgroups and found that the response inhibition ability of the children was the strongest predictor of the social approach behavior profiles. Similarly, Kirk, Kocking, Rigby, and Cornish (2013) found that heightened anxiety in some individuals with Williams syndrome was associated with attention allocation away from the eye regions of threatening facial expressions, whereas attention to eye gaze is typically overly fixated and prolonged.

This finding suggests that anxiety may play a role in biasing social attention in at least a subset of individuals with Williams syndrome.

In sum, persons with Williams syndrome show problems in social competence at the index level as well as an unusual pattern of intense social interest and attention to social stimuli coupled with social-emotional, social-communicative, and cognitive deficits. This profile of high social interest and poor social information processing likely contributes to the poor social outcomes (e.g., social isolation) but also makes these individuals particularly socially vulnerable in a variety of ways (e.g., bullying, abuse). In addition, despite relatively stable cognitive and linguistic functioning, and general improvements with age on adaptive skills, individuals with Williams syndrome remain dependent on family members and struggle with physical and mental health problems in adulthood.

Social Competence Among Persons With Prader-Willi Syndrome

Individuals with Prader-Willi syndrome eat excessively (may also ingest inedible items) and are typically morbidly obese. The pursuit of food is so extreme that in many cases they will resort to risk-taking and antisocial behavior such as lying, stealing, and exchanging sexual favors for the promise of food. These individuals exhibit poor social judgment and difficulty with interpreting social cues (Koenig, Klin, & Shultz, 2004). Rituals, tantrums, skin picking, and compulsive behaviors, such as hoarding and placing objects in a certain order, occur at elevated rates among persons with Prader-Willi compared with persons with other etiological syndromes (Greaves, Prince, Evans, & Charman, 2006; Holland et al., 2003). Clinically, the rates of autism and mood disorders are also elevated (Vogels et al., 2004).

Prader-Willi syndrome is caused by missing paternal chromosome 15 (q11–q13 region). Seventy percent of the cases are due to the deletion (DEL) of the region in the paternal contribution, 25% of the cases to the duplication of the region in the maternal contribution, referred to as uniparental disomy (mUPD), and the rest to either a methylation imprinting defect or a translocation microdeletion (Ledbetter et al., 1981; Nicholls et al., 1989). Individuals with the DEL subtype are more severely impacted than those with mUPD in several domains, including the characteristic facial appearance and hypopigmentation (Cassidy, 1984), maladaptive behaviors (Dykens et al., 1999), and self-injurious behaviors (Symons et al., 1999). However, individuals with mUPD are at increased risk for developing psychosis after

adolescence (Verhoeven & Tuinier, 2006) and autistic-like symptoms (Descheemaeker et al., 2006; Dimitropoulos & Schultz, 2007)

At the index level, based on parent ratings on the CBCL, children with Prader-Willi syndrome were reported to show poor behavior with others, and to be less active in social organizations compared with those with Down syndrome (Rosner et al., 2004). Their overall social competence did not improve with age as it did for children with Down syndrome and those with Williams syndrome (Rosner et al., 2004). Individuals with Prader-Willi syndrome also display poorer adaptive skills than would be expected given their intellectual functioning (Holland et al., 2003), and are reported by their caregivers to exhibit rigidity, inflexibility, and great difficulty with changes in their routines (Benarroch et al., 2007).

At the motivation/skills level, unusual but discrepant perceptual processing of faces was found in both genetic subtypes of Prader-Willi syndrome (Halit et al., 2008). In a preliminary neurophysiological study of individuals with Prader-Willi syndrome, an attenuated response to faces was found among eight individuals with paternal deletions (DEL), whereas the participants with mUPD showed a more typical neural response to faces although their eye gaze processing was similar to those participants with autism (Halit et al., 2008). Emotion recognition, a related area of social competence, is considered to be atypical in Prader-Willi, and may be related to high rates of comorbid affective psychosis (mUPD subtype) and clinical depression (DEL; Soni et al., 2008). Although Soni et al. (2008) found that caregivers drastically underestimated the emotion recognition and response abilities of both children and adults with Prader-Willi syndrome, problems in facial emotion recognition factors is thought to contribute to the peer relationship difficulties that are common among persons with Prader-Willi syndrome (Whittington & Holland, 2011). This is consistent with Tager-Flusberg et al.'s (1998) findings that adults with Prader-Willi syndrome performed significantly worse than adults with Williams syndrome when selecting the correct labels to match to the photographs of mental state expressions in the eye region.

Theory of mind also appears to be an area of impairment among persons with Prader-Willi syndrome. However, performance may vary with the type of task used or aspect of theory of mind that was measured. For example, Tager-Flusberg and Sullivan (2000) found that children with Prader-Willi syndrome (mean age 6 years, 11 months and mean IQ of 63) performed better on the false belief component of theory of mind than children with Williams Syndrome (mean age of 7 years 2 months

and mean IQ of 68). However, Koenig, Klin, and Schultz (2004) found that participants with Prader-Willi syndrome (mean age of 19.9 years and mean IQ of 67) performed as poorly as those with autism (mean age of 15.7 years and mean IQ of 69) and below IQ-matched participants without autism (mean age of 20.8 years and mean IQ of 72). The difficulties may be especially apparent when tasks require an appreciation for more abstract mental states as Koenig et al. used a social attribution task involving ambiguous stimuli (moving shapes in a video). In this context, the group with Prader-Willi syndrome made simple cognitive attributions about the moving shapes but were less able to make inferences about affective states related to the scenario (e.g., envy, jealousy), elements that were critical to understanding the social story.

Using within-group subtype analyses, Lo, Siemensma, Collin, and Hokken-Koelega (2013) found that both persons with the DEL and with mUPD subtypes showed poorer than expected performance on several components of theory of mind including first order belief, false belief, and second order false belief. In this group, 36% of children also met criteria for autism; 29% of the DEL and 41% of the mUPD subtypes. The most common symptoms of autism in the Prader-Willi syndrome group were in the area of maladaptive behaviors and routines such as interrupting conversations, talking to strangers, and making literal interpretations of expressions. Whereas most individuals with Prader-Willi syndrome do not exhibit the severity of symptoms found in autism, the social deficits that are reported appear to fall on the same continuum (e.g., poor social reciprocity and peer relationships).

Conclusions About Social Competence

The distinction between studies on index level, real-life summary indexes of social competence (e.g., peer acceptance, or employment success) and motivation/skills level, the underlying dispositions and abilities that provide the building blocks of social interactions (e.g., social motivations and social skills) is helpful to identify which aspects of social competence are impaired and the implications for overall adaptation. More of the research on social competence among the various etiological groups associated with intellectual disability has been focused at the motivational/skills level than at the index level and no studies were found that were focused on the relations between the foundational social abilities (e.g., face and emotion recognition) and real-life indexes of social competence (e.g., friendships). Understanding this relation will be informative about the extent to which current interventions, which are typically designed to tackle specific social

processes such as face and emotion perception can impact real-life social adaptation with all its complexity and many components. Thus, intervention studies designed to specifically target face or emotion processing in children with ID should also include outcome measures of indices of real-world social competence, such as peer and family relations.

Comparisons across syndromes point to unique profiles of social competence that further underscore the notion of multiple sciences of different etiologies rather than a single monolithic one in the study of persons with intellectual disability. Patterns of differences and similarities in sociability across syndromes would help us understand the contribution that sociability has on the overall social outcomes of persons with specific syndromes and provide further insight into gene–brain–behavior systems. For example, the hypersociability in Williams syndrome is clearly an extreme phenotype of a critical aspect of social competence. With increasing methodological sophistication, even more detailed analyses of within group subtypes and variation allows for more nuanced and precise genetic associations and markers with sociability. All of these extensions of the research must begin to be considered within a developmental framework to account for changes across syndromes across the lifespan rather than being based on extrapolations from studies with limited ranges of CAs or MAs.

Language Development

As with many areas of functioning, early accounts of language abilities among persons with intellectual disability were consistent with the defect theories. Various impairments were forwarded as characteristic of all persons with intellectual disability, regardless of etiology, and were generalized to involve all facets of language. This outlook has since evolved largely in relation to the emergence of the developmental approach to intellectual disability and the concordant increased specificity in the ability to measure different aspects of language. Accordingly, the discussion around language abilities among persons with intellectual disability has been transformed to include issues of development and abilities, rather than just impairment, and an emphasis on the etiology-specific developmental profiles of the many different components of language functioning. However, as with other areas of functioning, this more fine-grained and comprehensive approach only allows for the relatively extensive study of the most common etiological groups, and the intermittent study of a few others. Furthermore, statements about a general language skill

are misguided, as language is a multi-faceted process in which children often display multiple divergent trajectories that are largely dependent on foci of the research and the measures that are used.

To provide some insight into the role of language in understanding the whole person, we provide a brief review of current thinking regarding the disparate profiles of language processing as well as their interplay with cognitive and social development among the most commonly studied etiological groups, those of persons with fragile X syndrome, Down syndrome, and Williams syndrome. We also review language among persons with Prader-Willi syndrome as an example of current thinking in research on less studied syndromes. As with virtually all areas of functioning, the available evidence is piecemeal with only some areas of language functioning being considered within each etiological group, and with little consistency in the foci of research across groups.

Language Development Among Persons With Fragile X

Among children with fragile X, language is generally reported to develop along a consistent trajectory commensurate with MA, with the exception of pragmatics (Abbeduto et al., 2012). Individual differences within the syndrome are also reported as a function of gender, as males tend to be more affected overall than females, and as a function of autism comorbidity (which occurs in 20–30% of cases; Hatton et al., 2006; Rogers, Wehner, & Hagerman, 2001). In other aspects, inconsistent evidence leads to questions about language development in this group. In one example, Roberts et al. (2002) reported that receptive vocabulary lagged behind expressive vocabulary in a group of young boys whose MAs ranged from 12 to 28 months (CAs 21 to 77 months), whereas Abbeduto et al. (2003) reported that receptive vocabulary was at nonverbal MA expectations in a group of male and female adolescents. These findings suggest either that the rate of vocabulary acquisition increases more quickly than the rate of general cognitive development among individuals with fragile X over the course of development or that comparing receptive to expressive vocabulary (Roberts, 2002) leads to a different interpretation of strengths and weaknesses than comparing receptive vocabulary to MA (Abbeduto et al., 2003).

In a longitudinal study of boys with a mean MA of 5-years at entry, Martin, Losh, Estigarribia, Sideris, and Roberts (2013) found parallel gains on the Comprehensive Assessment of Spoken Language (Carrow-Woolfolk, 1999) among those with fragile X without autism, fragile X with autism, and Down syndrome over a 3-year period. In all

three groups, language appeared to be below non-verbal MA expectations, and both the boys with fragile X without autism and those with Down syndrome showed a pattern of vocabulary > pragmatics > syntax, whereas the boys with fragile X with autism showed a pattern of vocabulary > pragmatics = syntax due to poorer pragmatic performance in the comorbid autism group. Given the frequent reports of gender differences and consequential exclusion of females from many studies, the pattern of findings reported by Abbeduto et al. (2003), which were inconsistent with other studies, might be related to the inclusion of females in their study (Abbeduto et al., 2012). This was supported by findings from a longitudinal study of boys and girls with fragile X, in which Pierpont, Richmond, Abbeduto, Kover, and Brown (2011) found that phonological and verbal working memory predicted gains in vocabulary and syntax for boys, but not girls, with fragile X. The evidence from the study of language among individuals with fragile X highlights the importance of taking into consideration both gender and the presence of a comorbid diagnosis of an autism spectrum disorder in understanding the development of skills in this group.

Language Development Among Persons With Down Syndrome

In a review of language development from childhood to young adulthood among persons with Down syndrome, Chapman and Kay-Raining Bird (2012) note that children with Down syndrome display a specific phenotype of language learning, including uneven impairments in language skills, yet they are one of few etiological groups in which impairments in pragmatics (the social use of language) are not identified. Combining evidence from cross-sectional and longitudinal studies of language acquisition, Chapman and Kay-Raining Bird (2012) painted a picture of changes in language functioning across development. In early childhood, children with Down syndrome display deficits in expressive vocabulary that are below nonverbal MA expectations (Fidler, Hepburn & Rogers, 2006; Miller, 1995). However, from age 2 to 4 years, the rate of expressive vocabulary growth falls further and further behind both receptive vocabulary knowledge and nonverbal development (Miller, 1995), with receptive vocabulary remaining commensurate with non-verbal development until early adolescence (Chapman, Schwartz, & Kay-Raining Bird, 1991). As the grammatical complexity of language skills among typically developing children increases, children with Down syndrome appear to fall further and further behind, even as language skills continue to grow (albeit unevenly).

In an attempt to study the relationship between language and cognition among individuals with Down syndrome, Chapman, Hesketh, and Kistler (2002) conducted a 6-year longitudinal study of grammar in which they found that receptive syntax at the beginning of the study predicted later expressive syntax. Furthermore, short-term memory skills (both auditory and visual) predicted the rate of receptive syntax growth, which in turn predicted the rate of growth in Mean Length Utterance. Auditory short-term memory has also been implicated in phonological development (Kumin, Councill, & Goodman, 1994; Laws & Gunn, 2004), the emergence of which appears to be delayed among infants with Down syndrome, although the pattern has been reported to follow a typical trajectory (Smith & Stoel-Gammon, 1983).

Consistent with the developmental approach, Chapman and Kay-Raining Bird (2012) caution that the manner in which we measure development plays a role in our understanding of language-related strengths and weaknesses of individuals with Down syndrome, and could lead to an under-evaluation of abilities. For example, researchers debate the relative strength of receptive vocabulary among individuals with Down syndrome during adolescence, with Chapman et al. (1991) reporting that receptive vocabulary surpasses general MA, and others reporting that receptive vocabulary is below MA expectations (Hick, Botting, & Conti-Ramsden, 2005; Price, Roberts, Vandergrift, & Martin, 2007; Roberts, Hennon, et al., 2007). However, this discrepancy in findings may have more to do with the use of different MA measures across studies, rather than actually reflecting conflicting patterns of findings. Further, with respect to grammar, when assessments of language are based on conversational samples (e.g., question–answer or spontaneous comments during free play), expressive syntax appears to reach a plateau in adolescence among individuals with Down syndrome (Chapman et al., 1998; Fowler, Gelman, & Gleitman, 1994). Conversely, when assessments of language are based on narrative samples, the development of expressive syntax continues through young adulthood (Chapman et al., 1998; Chapman et al., 2002; Thordardottir, Chapman, & Wagner, 2002).

Language Development Among Persons With Williams Syndrome

Some early accounts of individuals with Williams syndrome focused on apparent strengths in expressive language, to the extent that in an early account, Bellugi, Marks, Bihrlé, and Sabo (1988) described them as demonstrating a double dissociation between language and cognition or as having intact language abilities despite

a general level of functioning in the range of intellectual disability. However, Mervis (2012) and Karmiloff-Smith (1998, 2007) argue that the expressive language abilities of individuals with Williams syndrome only appear to be so outstanding when they are considered in relation to the extremely impaired skills, such as visuo-spatial construction, in this group. With closer scrutiny, various aspects of language also appear to be delayed or impaired, and the development of language appears to be interdependent with, rather than independent from, general cognitive development (Mervis, 2012).

Karmiloff-Smith et al. (1997) argue that the entire trajectory of language development is altered in Williams syndrome, and that this may be linked to the syndrome's characteristic hypersociability. Beginning in infancy, infants with Williams syndrome seem to pay excessive attention to people at the expense of joint attention behaviors, which are necessary precursors to typical language development (Bloom, 2002). Despite, or perhaps consequential to, their hypersociability, atypicalities have been reported in virtually all areas of pragmatic language abilities, which include language related to social interaction such as turn taking, modulating content for the audience, and communicating wants and needs (Philofsky, Fidler, & Hepburn, 2007). Specifically, individuals with Williams syndrome exhibit a particular pattern of unusual conversation that tends to include features such as excessive idioms, overfamiliarity, evaluative comments, and emphatic markers, a cocktail party speech that is partly responsible for the reputation of strong language (Mervis, 2012). Individuals with Williams syndrome are quite verbose, but this verbosity appears to mask difficulties in the social aspects of language, the very skill that was initially considered to be their primary strength.

Language Development Among Persons With Prader-Willi Syndrome

The difficulties in providing a comprehensive understanding of less common etiological groups are highlighted in the literature on the development of language among individuals with Prader-Willi syndrome. Impairments in both expressive and receptive language have been cited, with greater impairment reported in expressive language (Branson, 1981; Dimitropoulos, Ferranti, & Lemler, 2013; Kleppe, Katayama, Shipley, & Foushee, 1990; Munson-Davis, 1988), although these findings are plagued by methodological problems regarding the comparison groups. For example, findings that expressive language appears to be particularly problematic in individuals with Prader-Willi syndrome, with specific problems with

morphosyntax and possible difficulties with vocabulary and pragmatics (Van Borsal, Defloor, & Curfs, 2006) are compromised as they are framed only in relation to the expected norms for CA. Although preliminary, areas of strength in language abilities among individuals with Prader-Willi syndrome have also been suggested in phonology (Van Borsal et al., 2006) as well as in vocabulary knowledge and decoding in the domain of reading (Dykens, Hodapp, Walsh, & Nash, 1992).

The differentiation among subgroups within Prader-Willi provides an opportunity to better understand the relation between aspects of the disorder and those of language abilities, although the evidence is mixed. For example, Dimitropoulos et al. (2013) reported that participants with maternal uniparental disomy (mUPD) genetic subtype of Prader-Willi syndrome only show higher expressive language abilities relative to their receptive abilities, whereas Lewis, Freebairn, Heeger, and Cassidy (2002) reported no differences in speech or language between individuals with Prader-Willi syndrome from either deletion or maternal uniparental disomy. If researchers are able to cobble together a more comprehensive literature on language processing among persons with Prader-Willi despite the difficulties, the subtle differences between subtypes may be essential to understanding the genetic mechanisms of intellectual disability in this population and their relationships to language abilities.

Language, Cognition, and Socialization

Although some language skills appear to develop independently of other cognitive abilities in some etiologies, the interdependence of language, cognition, and socialization is evident (Chapman & Kay-Raining Bird, 2012), albeit in different ways across groups. For example, Mervis (2012) notes that intact verbal working memory is important for grammatical comprehension among children with Williams syndrome, more so than for typically developing children. Among individuals with Down syndrome, Kay-Raining Bird and Chapman (2012) highlight the interdependence of language in literacy development, as vocabulary comprehension is reported to be the best predictor of their literacy skills (Kay-Raining Bird, Cleave, White, Pike, & Helmky, 2008).

Syndrome-specific patterns in language development may also play a role in the acquisition of other higher order cognitive skills, such as executive functions. For example, Campbell et al. (2013) found that verbal development, independent of nonverbal development, predicted cognitive flexibility among persons with Down syndrome (Campbell et al., 2013), and Landry, Russo, Dawkins,

Zelazo, & Burack (2012) found that it predicted working memory as well as cognitive flexibility among both persons with Down syndrome and those with Williams syndrome. This type of quest to understand the interrelatedness among skills across development exemplifies an essential task of the developmental approach, but is not sufficient. Rather, in addition to psychologically measurable intrinsic factors which impact the development of language, the complexity of genetic and environmental factors and their interplay must be considered in relation to the development of unique profiles of language, or any other type of, skills specific to the etiological groups. This was highlighted by Kay-Raining Bird and Chapman (2012) who describe how school, SES, and the home environment can affect the development of literacy skills among children with Down syndrome. For example, individuals with Down syndrome raised in a home environment display higher reading scores than those raised in an institution (Bochner, Outhred, & Pieterse, 2001; Carr, 2000), and those in integrated school settings and from higher SES backgrounds also display better literacy skills (Bochner et al., 2001; Sloper, Cunningham, Turner, & Knussen, 1990).

In delineating that genetics must also be considered when studying the relations among socialization, the environment, and language abilities, Abbeduto et al. (2012) reviewed evidence of links between language skills in fragile X and levels of FRMP, the protein produced by the FMR1 gene that plays a critical role in synapse maturation and functioning. Specifically, levels of FRMP are reported to be related to communication scores, verbal IQ, and story comprehension (Bailey, Hatton, Tassone, Skinner, & Taylor, 2001; Kuo, Reiss, Freund, & Huffman, 2002; Simon, Keenan, Pennington, Taylor, & Hagerman, 2001). Yet interactions with parents also affect the development of language skills in children with fragile X. For example, maternal responsivity is reported to be associated with communication in children with fragile X (Warren, Brady, Sterling, Fleming, & Marquis, 2010). Specifically, Warren et al. (2010) reported that level of maternal responsivity was related to the rate of total communication, receptive and expressive language development in 36-month-old infants with fragile X syndrome. These types of findings regarding the influence of parents underscore Abbeduto et al.'s call for an increased consideration of the complex role of genetics in language development. For example, mother carriers with the full or premutation of FMR1 gene may themselves experience cognitive deficits or affective disorders, which may inadvertently contribute detrimentally to their children's language development (Abbeduto et al., 2012).

The Impact of a Child With Intellectual Disability on the Family

Consistent with the notion of understanding the whole person, the study of aspects of development that are intrinsic to the individual need to be supplements with regard to the study of family relationships. At every moment, the unique development of the child influences the signals that he or she sends out to the world. Beginning with the simple characteristics in temperament, the child demands more or less attention from the caregiver, which may be easily accommodated or may stretch the limits of the caregiver. The caregiver then approaches the child with a positivity or negativity that is in part influenced by the child. For example, consistent with the notion that children with Down syndrome have the reputation of being easy babies, the caregivers of children with Down syndrome report lower levels of stress than caregivers of children with other diagnoses such as autism (Dabrowska & Pisula, 2010) and Prader-Willi syndrome (Lanfranchi & Vianello, 2012). This easier personality likely leads to more positive interactions with parents and siblings.

These findings highlight that, as with all persons, persons with intellectual disability both affect and are affected by the close relationships in their lives, such as those with family members. Clearly, having a child with intellectual disability can uniquely impact the well-being of parents, siblings, and the family as a whole. Just as obviously, different etiologies can be associated differentially with family well-being since differences in development and symptomology may exert different types of stressors on parents and may also affect the availability of certain resources or coping mechanisms. This effect is transactional, since the impact on parental, sibling, and family well-being continually shape parenting techniques and other factors which are, in turn, impacting the well-being of the child, all within the context of the larger, community, and society in which the child and family reside.

Negative Emotional Outcomes for Parents of Children With Intellectual Disability

Not surprisingly, having a child with intellectual disability can impose a negative emotional toll on parents. Depression has been the most common negative outcome studied in parents of children with intellectual disability (Glidden, 2012), which appears to diminish with time. High rates of parental depression are more often reported soon after the birth of a child with intellectual disability relative to years later during childhood and adolescence (Glidden & Jobe, 2006; Glidden & Schoolcraft, 2003;

Keogh, Garnier, Bernheimer, & Gallimore, 2000; Singer, 2006). Anxiety, pessimism, and anger have also been reported (Glidden, 2012; Hall, Bobrow, & Marteau, 2000; Hodapp, Dykens, & Masino, 1997). These reactions are often in reaction to specific aspects of the child's specific condition. For example, increases in pessimism in parents are correlated with increases in maladaptive behavior in children with Smith-Magenis syndrome (Hodapp, Fidler, & Smith, 1998) and with Prader-Willi syndrome (Hodapp, Dykens, & Masino, 1997).

In addition to the negative emotional impact that having a child with intellectual disability has on the parents individually, it has also been reported to negatively affect the parental dyad (Glidden, 2012). Having a child with intellectual disability has been reported to slightly decrease marital adjustment (Risdal & Singer, 2004) and increase divorce rate (Reichman, Corman, & Noonan, 2004), although Hodapp (2007) reported a lower divorce rate for parents of children with Down syndrome relative to parents of typically developing children thereby indicating that the specific developmental profile of children with Down syndrome may uniquely protect against negative marital outcomes. Marital outcomes among parents of children with intellectual disability have been reported to be related to paternal involvement (Simmerman, Blancher, & Baker, 2001; Willoughby & Glidden, 1995), with more marital satisfaction reported in couples in which the father is involved in caring for the child (Willoughby & Glidden, 1995) or at least perceived by the mother as being involved (Simmerman, Blancher, & Baker, 2001). Coping style is also related to marital satisfaction among parents of children with intellectual disability. For example, the use of problem-focused coping by both parents in dealing with hassles has also been found to be related to higher marital adjustment (Stoneman & Gavidia-Payne, 2006).

Positive Outcomes for Parents of Children With Intellectual Disability

Despite the challenges often associated with raising a child with intellectual disability, many of the same positive life changes and emotions such as love, joy, and satisfaction that are experienced by parents of typically developing children are also reported anecdotally and in qualitative reports by parents of children with intellectual disability (see Hastings & Taunt, 2002). For example, Flahery and Glidden (2000) reported that lower levels of depression, positive marital adjustment, and increased family strength were found in both adoptive and non-adoptive parents of children with intellectual disability. In addition, mothers and fathers who felt optimism, contentment, and happiness

also had less stress and better family adjustment (Trute & Hiebert-Murphy, 2005).

Predictors of Parental Outcome

The stress of receiving a diagnosis is one factor often reported to elicit negative reactions and elicit symptoms of depression in parents soon after the birth of a child with intellectual disability (Glidden & Jobe, 2006, 2009; Glidden & Schoolcraft, 2003; Poehlmann, Clements, Abbeduto, & Farsad, 2005). Although this certainly can take an emotional toll, not receiving a specific diagnosis can be just as distressing for a family with a child who is not developing at the same rate as his/her peers (Glidden, 2012). A difference in the timing of when parents receive their child's diagnosis is thus one factor that has been reported to affect positive or negative outcomes depending on the etiology of the intellectual disability (Lenhard, Breitenback, Ebert, Schindelbauer-Deutscher, & Henn, 2005; Poehlmann et al., 2005). Specifically, parents report less anxiety, worry, and regret (Lenhard et al., 2005), and even relief (Poehlmann et al., 2005) when their children receive a diagnosis. Concordantly, Al-Yagon and Margalit (2012) highlight that early knowledge of a diagnosis was related to better parental adjustment among parents of children with Down syndrome. This is exemplified by findings that parents of children with Down syndrome generally show levels of anxiety, depression, and parental stress that are similar to those of parents of typically developing children (Hall, Bobrow, & Marteau, 2000), but not when they received a false negative diagnosis during prenatal screening (Hall & Marteau, 2003). In those situations, they display high levels of parenting stress, a negative attitude toward their child, and worse overall adjustment.

The type of coping mechanisms that are used and their availability to the parents also have been found to predict family well-being. Coping strategies that are focused more on problem solving and social support are related to more positive emotional outcomes than those using denial escape or avoidance of difficulties (Altshuler & Ruble, 1989; Levy-Shiff, Dimitrovsky, Shulman, & Har-Even, 1998; Reichman, Miller, Gordon, & Hendricks-Munoz, 2000). For example, mothers who use escape avoidance coping have been found to report higher levels of depression, whereas mothers who employ positive reappraisal report higher levels of subjective well-being (Glidden et al., 2006). Gender differences in coping styles have also been reported, as mothers tend to seek more emotional support whereas fathers seek more instrumental support and problem solving (Sullivan, 2002).

The children themselves, and especially their behaviors, impact considerably on parental well-being (Glidden 2012), as increases in child behavior problems are reported to contribute to more negative parental outcomes than the diagnosis of intellectual disability itself (Baker et al., 2002, 2003). Consistent with the findings that parents of children with Down syndrome experience a relative advantage as compared with parents of children with other etiologies, Al-Yagon and Margalit (2012) suggest that less severe behavior problems in children with Down syndrome may contribute to more positive outcomes for their families since less stress is associated with getting such behaviors under control.

Dyadic Interaction Between Mothers and Children With Intellectual Disability

One contributor to the parents' level of well-being that is affected by the child with intellectual disability is the parent-child dyad with that child. As with other aspects of social development and the family structure, the impact must be considered in relation to the relevant construct, task, and etiology (Hauser-Cram, Howell-Moneta, & Mercer-Young, 2012). In one example of an etiology-specific finding, Vaughn, Goldberg, Atkinson, Marcovitch, MacGregor, and Seifer (1994) found that young children with Down syndrome display an unclassified insecure attachment style relative to typically developing children who generally show a secure attachment to their mothers, as they are less reactive to their mothers and seek their mothers less for reassurance upon return. Esbensen et al. (2012) suggest that, in addition to deficits in cognitive functioning, the increase in the unclassified insecure attachment style among children with Down syndrome relative to typically developing children may be due to etiology-specific characteristics of Down syndrome that put strain on the mother-child relationship. Characteristics such as congenital heart defects, thyroid dysfunction and sleep problems, and ear infections can all limit the energy of children with Down syndrome and their ability to engage in sustained interactions with their mothers. Children with Williams syndrome display a similar attachment style as they show less intense facial and vocal distress when their parents leave the room and need less consoling upon being reunited with parents (Jones et al., 2000).

Mother-child interactions illustrate the etiology specific effects on parenting style. For example, parents of children with Williams syndrome were found to use more task directives than parents of children with Prader-Willi syndrome when playing with a puzzle, a style apparently adopted due to their children's poorer spatial skills (Ly &

Hodapp, 2005). In addition, the unique hypersocial personality style of children with Williams syndrome can be both beneficial and detrimental to the relationship (Jones et al., 2000). Accurate recognition of positive emotions creates a synergy of positive bi-directional affects in the dyad, although extensive interactions with others may diminish the child's interactions and relationship with their own mother (Järvinen-Pasley et al., 2008; Laing et al., 2002). With regard to Down syndrome, infants and toddlers appear to show less coordinated shared attention with their mothers as compared with typically developing children matched on MA (Legerstee & Fisher, 2008).

In keeping with Zigler's (1967; Zigler & Hodapp, 1986) argument for considering the ways that being a person with intellectual disability inevitably involves life experiences that affect the ways that people behave and perform in all aspects of their lives, the areas of social competence, language development, and family relationships are all essential to real lives lived. The simple cataloguing of the phenotypes as though the genotype is the only contributing factor to the eventual developmental path is clearly an erroneous strategy as features of different phenotypes present particular challenges for optimal functioning and relating within the context of the whole person in his or her family and environment.

NEUROSCIENCE AND THE DEVELOPMENTAL APPROACH: BENEFITS AND PITFALLS IN THE APPLICATION OF CUTTING-EDGE TECHNOLOGY

The advance of technology and the increase in the number of researchers who have access to cutting-edge tools like functional magnetic resonance imaging (fMRI), event-related potentials (ERPs), and magnetoencephalography (MEG) have led to a growing number of studies that involve mapping the relations between how an individual performs on a task and the brain regions that support that performance. The potential utility of these increasingly sophisticated and precise technologies along with the concomitant theoretical and methodological contributions of the field of neuroscience provide promise for yet a new stage in the development of the developmental approach to the study of persons with intellectual disability. Concordantly, as neuroscientists begin to delve into the study of intellectual disability, the history of the developmental approach can guide them in detecting theoretical and methodological landmines to avoid and directions of promise to pursue.

A Primer on What fMRI and ERP Measure

The study of brain function, which reflects modern techniques for linking behavior to brain, has generally involved either fMRI or ERP technology. Briefly, fMRI is based on the basic principle that any given region of the brain that is actively engaged in a task will draw more blood. This blood, rich in oxygen, is detected magnetically due to its iron content. fMRI is used to measure the so-called blood-oxygenated dependent level (BOLD) response, which serves as an indirect marker of neuronal activation during a cognitive operation. Namely, neural firing increases in a particular brain area as it is being engaged, which leads to both greater blood flow and release of oxygen to this area to satisfy local metabolic demands. By contrasting experimental conditions that differ only in the cognitive construct one is measuring or by comparing task based activations to rest, researchers can isolate the location of brain activity specific to a particular task or cognitive process. The major advantage of fMRI is that it has exquisite spatial resolution, allowing scientists to locate the specific areas of the brain that underlie task performance with millimeter precision. One of the disadvantages is that, because blood flow changes occur slowly, the temporal resolution of fMRI is on the order of seconds, which is much slower than the timing of brain processing in general, which is in the order of milliseconds.

In contrast to fMRI, event-related potentials (ERPs), offers the fine-grained temporal resolution researchers seek, but at the expense of spatial resolution. In order to relate brain activity to specific processes and behaviors, researchers isolate the continuous, ongoing electrical activity of the brain as measured by encephalography (EEG) into segments that are time-locked to a specific stimulus (Luck, 2005). ERPs display a stable time relationship to a discrete event (Luck, 2005). Because ERPs are small in size relative to other physiological events, many presentations of the evoking stimuli are necessary to average out the potentially unrelated events (Luck, 2005). For example, if a tone is played, the brain responds to that tone, but it also responds to other things that are present in the environment at that same time such as how a person is feeling, what they are looking at, and what they are thinking about. By time locking the physiological response to the presentation of the tone, and presenting the tone repeatedly, we can reduce the noise (the brain's response to things other than the tone) and extrapolate the brain's specific response to the tone itself (Luck, 2005). The brain's responses to a stimulus generate an ERP response that has specific peaks and troughs whose amplitude (height)

and latency (timing) can be compared either between conditions or between groups. The nomenclature of ERPs is based on the direction (positive or negative from 0) and time post stimulus onset, such that a P100 (or P1) represents a positive peak occurring 100ms post stimulus and an N170 represents a negative peak that occurs 170ms post stimulus. The meaning of each of these components have been isolated (with considerable disagreement) to reflect perceptual, attentional, or cognitive mechanisms and the stimulus factors to which they are sensitive.

An ERP represents only a sample of the electrical activity that is present in the brain when a specific event occurs (Luck, 2005). Since electrodes are located at the surface of the scalp, the activity of one single neuron is too small (and too far away) to record. Thus, ERPs are used to measure the integrated activity of many neurons that are recorded at the surface of the scalp. Specifically, neurons must have their dendritic trees oriented to one side (Coles, Gratton, Kramer & Miller, 1986) to be measured. ERPs are generally thought to reflect postsynaptic potentials (Luck, 2005) because these occur in the dendrites essentially instantaneously, while action potentials need to travel down the axon of a neuron (Coles et al., 1986; Luck, 2005). The advantage of this technique is that its temporal resolution is in the order of milliseconds, which reflects the speed with which our brain processes information. This allows researchers to examine different levels of processing such as perception and cognition by looking at early (first 250ms or so) and late (after 300ms) time frames of processing, which can reflect afferent (receiving input) or efferent (sending output) neural pathways. The disadvantage of ERPs is that because electrodes are placed at the surface of the scalp, which is far from the brain, the spatial resolution of ERPs is poor. Thus, fMRI is best suited to tell us *where* in the brain things happen, whereas ERPs allow us to know *when* things happen.

Neuroscience and the Developmental Approach: A Messy Meeting of Disciplines

The lessons learned from the developmental approach to intellectual disability which are particularly relevant to the emergence of the study of intellectual disability from a neuroscience perspective relate primarily to matching issues. One, comparing individuals with an intellectual disability to typically developing individuals who have normal cognitive function is problematic because it confounds general cognitive deficits with any specific skill or process being measured. Two, when matching on the basis of MA, the composition of the comparison group is

particularly important and should be homogeneous (e.g., Down syndrome) as opposed to mixed (i.e., participants from different groups) such that (1) direct comparisons can be made between two groups that differ in only the cause of their intellectual disability, and (2) researchers can replicate findings by having similarly composed comparison groups. As will become evident from the review of studies that follow, these lessons, which form the basic tenets of the developmental approach to intellectual disability, are often ignored in studies of intellectual disability in which fMRI or ERP technology are used.

Generally, when reviewing studies on intellectual disability within the context of the developmental approach, papers in which the performance of individuals with intellectual disability are matched to the CAs of typically developing participants are discounted since performance differences are simply attributable to the a priori differences in level of functioning between the groups, reflecting the defect, rather than the developmental, approach. However, the notion of how best to apply the developmental approach to the study of brain processes still needs to be considered. For example, in the absence of a cognitively demanding task in which age-related differences would be expected, do we expect the neuroanatomical substrates (in the case of fMRI) or the neurophysiological components (in the case of ERPs) engaged or elicited by the presence of a stimulus to reflect physical or mental maturity? Would patterns of brain activation of a 12-year-old individual with Down syndrome whose MA is 6 resemble that of a 12-year-old or that of a 6-year-old, or somewhere in between? Does this vary as a function of the level of intellectual impairment? Is the answer to this question diagnosis-dependent? Task dependent? Answers to these questions will help to define and refine the reciprocal relationship, not only between typical and atypical development but also between developmental psychopathology and neuroscience.

Neuroscience and Intellectual Disability

As applied to the study of intellectual disability, the study of neurophysiology and neuroimaging are in their infancy. In applying the developmental approach to assessing the contributions of neuroscience (broadly speaking) research in the study of intellectual disability, we review all of the research studies that we could find in which functional magnetic resonance imaging or event-related potentials were the primary methodological tool. Consistent with the developmental approach, the discussion of these articles will be focused on both the main findings from these studies and how they contribute to our understanding of both

persons with intellectual disability and of typically developing persons, but also the implications of matching or non-matching practices for these findings and subsequently on advancing the field. Studies were selected for review according to the following course of action. An advanced PubMed search of the most common genetic disorders associated with intellectual disability—Down syndrome, Williams syndrome, fragile X, and Prader-Willi—was conducted with either the terms *event-related potentials* or *functional magnetic resonance imaging*. In addition, we looked through the reference sections of the articles generated through our original search and added additional relevant articles. This extended search yielded a total of 49 published research studies, 35 of which were studies that involved fMRI technology and 14 that involved ERPs as their main methodological tool. Abstracts of these studies were then read to exclude reviews and other non-empirical papers ($n = 6$), or papers that were irrelevant to perception, attention, and cognition very broadly defined (e.g., articles whose primary focus was on Alzheimer's disorder among persons with Down syndrome, or those looking at resting state fMRI were rejected; $n = 12$), or those for which full text articles could not be located ($n = 3$). This left a total of 28 articles that are reviewed here. The details of each study, including the chronological and mental ages of participants, and the matching strategies used by researchers are presented in Table 1.1, and are sorted as a function of etiology. Though the matching strategies used by researchers will be highlighted in the text, we refer the reader to the table for the specific participant and matching characteristics. Also included in the table are the relevant methodological factors, and the main research findings.

As the application of neuroscience techniques to the field of intellectual disability is nascent, we found little consistency in findings, or even in areas of study. The themes of sociability and food-related preferences represent the majority of areas of inquiry in the study of individuals with Williams and Prader-Willi syndromes respectively as these areas represent such consistent aspects of the phenotype for both of these disorders. However, no such consistency is to be noted across other areas of intellectual disability. Accordingly, few studies are replicated and no firm conclusions can be drawn regarding areas of strength or weakness or areas of the brain that are over or under active in response to specific types of stimuli. Although the main findings across studies will be reviewed, the main focus of this review is on the relationship between findings and the developmental approach to intellectual disability as it applies to neuroscience.

Matching in Neuroscience Studies. When reviewing the different studies, three broad matching classifications were used. The first category included studies with CA-matching alone. The majority of the studies ($n = 19$) fell under this umbrella of problematic methodology. The second category of studies ($n = 8$) involved the consideration of developmental level either directly with matching mental age or statistically by covarying IQ in the analyses. Finally, one study involved matching on a specific aspect of functioning (temperament) to assess the specificity of social approach among persons with Williams syndrome.

The publication of these papers in high-impact journals, despite the omission of any mention of such foundational issues in the developmental approach as the role of discrepant IQs or level of functioning, is particularly troubling. Conversely, many developmentalists are beginning to conduct research using cutting-edge technology without the necessary background training. As such, basic conventions in the ERP and fMRI literatures regarding methodology, such as compensating for multiple comparisons in fMRI and testing for assumptions underlying statistical tests such as sphericity in ERP data, which are common knowledge among basic neuroscientists in the field and essential to the acceptance for publication are often overlooked. One explanation is that due to the amount of effort required to develop meaningful tasks that can be completed by atypical populations and the difficulty involved in recruitment and testing, some of the methodological rigor of basic science papers gets overlooked (for information about basic considerations in fMRI research, see Bennett, Wolford, & Miller, 2009; Bennett, Baird, Miller, & Wolford, 2010; for considerations related to ERPs, see Picton et al., 2000). We review the available etiology-specific neuroscience-type studies within the context of the basic fundamentals of both the developmental approach and cutting-edge technologies.

Persons With Fragile X Syndrome. Due to the gender differences associated with the presence of an unaffected, protective X chromosome among females with fragile X syndrome, males and females are generally studied separately. Differences in their cognitive phenotypes are commonly associated with the lack of intellectual impairments noted among females with fragile X. We identified 10 neuroscience-based studies of fragile X, half of which involved fMRI to assess various aspects of functioning that included cognitive interference via the Stroop task, auditory discrimination, and emotional identification among females with fragile X, and the detection of eye gaze direction and inhibition using a go/no-go task among males, and

TABLE 1.1 Summary of Studies Using Neuroscience Tools Among Individuals With Developmental Disabilities

| Citation | Diagnosis and comparison group (<i>N</i> , age, IQ)* | Matching* | EEG or fMRI | Stimuli and task | Findings (RT = Reaction time) |
|-----------------------|--|--|-------------|---|--|
| Watson et al. (2008) | <p>FXS: <i>N</i> = 13. Age: <i>M</i> = 15.5, <i>SD</i> = 2.4. IQ: WISC III: <i>M</i> = 61, <i>SD</i> = 14.8. DD (developmental delay) group: <i>N</i> = 13. <i>M</i> = 16.1; <i>SD</i> = 3.3. IQ <i>M</i> = 62.4, <i>SD</i> = 9.4. TD: <i>N</i> = 13. Age: <i>M</i> = 15.0, <i>SD</i> = 2.5. IQ: <i>M</i> = 116.8, <i>SD</i> = 11.6.</p> | <p>CA matched to TD, IQ and CA matched to DD</p> | fMRI | <p>Color photos of faces with neutral expressions that were (1) forward facing with a direct gaze, (2) forward facing with averted gaze, (3) averted face with direct gaze, and (4) averted face with averted gaze. Button press for averted or direct gaze.</p> | <p>Aberrant neural processing during gaze perception and a lack of neural adaptation to successive gaze stimuli were found in the group with FXS. Functional brain correlates related to a decreased ability to adapt to direct eye gaze stimuli. Behavioral data: Task accuracy differed between the TD group and other groups (but not between the DD and FXS groups). fMRI Data: Within-group comparisons of the contrast DirectGz-AvertedGz revealed significant activation in distinct regions: (1) FXS group showed significant activation in the left anterior insula. (2) TD group in the right parahippocampal gyrus and the left medial frontal gyrus. (3) DD group in the right midfrontal and parahippocampal gyri. Whole-brain ANOVA for the DirectGz vs. AvertedGz contrast revealed a main effect of group in the right cingulate gyrus, right midfrontal gyrus, and left insula. ROI analyses: main effects of group in the left amygdala and in the right STG. FXS had significant sensitization in the left amygdala which correlated negatively with performance on successive trials in the FXS group.</p> |
| Garrett et al. (2004) | <p>FXS: <i>N</i> = 15 but 4 discarded; all female. Age: <i>M</i> = 16.4, <i>SD</i> = 4.09, Range = 10–22; IQ: (WISC III for children, WAIS II for adults); <i>M</i> = 93.7, <i>SD</i> = 10.4. TD: <i>N</i> = 15 all female. Age: <i>M</i> = 15.5, <i>SD</i> = 3.4, Range = 10–22. IQ: <i>M</i> = 107.0, <i>SD</i> = 11.2). TD group had higher full-scale IQ score than FXS.</p> | <p>IQ matched and gender</p> | fMRI | <p>Color photographs of faces in four categories: (1) face forward with direct gaze, (2) face forward with averted gaze, (3) face angled with direct gaze, (4) face angled with averted gaze. Task 1: Subjects pressed a button if the face in the photograph was looking at them or another button if the person was looking away from them. Task 2: Subjects were required to indicate direction of gaze for faces and to alternate pressing the first and second button in response to the scrambled pictures.</p> | <p>Results included 11 subjects in the FXS group and 11 subjects in the TD. (1) Task Accuracy: TD had greater accuracy (2) Full scale IQ: Scores were correlated with task accuracy for both groups combined but not within each of the groups. Subsequent between-group analyses were covaried for IQ, which also controlled for between-group differences in the task accuracy. (3) RT: No group differences. (4) Brain activation (direct-averted faces): No differences comparing forward faces with angled faces. Brain activation (direct-averted gaze)—Three clusters of greater activation were noted in the TD group including STS, lingual gyrus and cerebellum. Two clusters of greater activation were noted in FXS relative to TD group including the right insula and the cerebellum. Activation FG did not differ between FXS and TD. Interaction between group and hemisphere showed TD right > left FG activation to all stimuli.</p> |

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| Hall et al. (2009) | <p>FXS: $N = 10$, females; Age: ($M = 18.7$ years, $SD = 3.81$ years). IQ (test not specified) $M = 86.3$, $SD = 16.41$. TD: $N = 10$, female; Age ($M = 14.7$ years, $SD = 2.95$ years). IQ ($M = 110.2$, $SD = 10.84$)</p> | MA matched. | fMRI | Two tones (standard and comparison): determine by button press whether the comparison tone was longer or shorter than the standard tone. In control task the stimuli were of equal duration. | <p>Accuracy and RT did not differ between the groups. Brain Activation: FXS activation was more diffuse and distributed over a large number of regions. No greater activation in TD compared with FXS. IQ and age—no correlation with activated brain regions; in FXS only the left pons showed a significant (positive) correlation with mental age. Percent correct correlated with activation in the cerebellum in the TD group, and performance was significantly correlated with activation in middle temporal gyrus in the FXS group. In the FXS group, no significant correlation with FMRP level.</p> |
| Hagan et al. (2008) | <p>FXS: $N = 10$, female. Age: $M = 16.4$, $SD = 4.9$, range = 9.7–24.0 years. IQ: (WISC-III for children, WAIS III for adults) $M = 91$, $SD = 16.2$, range 75–124; TD: $N = 10$, female. Age: $M = 15.6$, $SD = 4.2$, Range = 8.4–22.9; IQ: $M = 106.1$, $SD = 15.7$, range 79–128. No age differences; trend for FSIQ differences between groups.</p> | CA matched. | fMRI | Color photographs: happy, sad, neutral and scrambled faces. Press the left button if the person in the photograph appeared happy, a middle button if the person appeared sad, and a right button if a neutral or scrambled face appeared. | <p>Accuracy: FXS participants were less accurate at recognizing neutral and scrambled faces relative to TD. No differences in the identification of happy or sad faces. RT: no differences. No correlation for either group between RT and FSIQ. Brain activation: increased left hemisphere activation in left medial frontal gyrus and left superior and middle temporal gyrus, left cerebellum and left pons among participants with FXS relative to TD. Overall more activation in FXS than TD.</p> |
| Hoefl et al. (2007) | <p>FXS: $N = 10$, all male. Age: $M = 15.4$, $SD = 2.7$; IQ (WISC-III for children, WAIS III for adult): $M = 59.2$, $SD = 9.8$. Idiopathic (DD)- $N = 10$, male. Age: $M = 14.6$, $SD = 2.7$; IQ: $M = 65.3$, $SD = 13.8$. TD: $N = 10$, male. Age: $M = 16.7$, $SD = 4.2$; IQ: $M = 125.6$, $SD = 11.5$. The three groups were matched for age and race/ethnicity. IQ of TD > DD = FXS</p> | FXS and DD matched on IQ; FXS, DD, TD were matched for age and race/ethnicity. | fMRI | Visual go/no-go task: Press the button to every letter except X which occurred 50% of the time; Control task—press a button to every letter but no x's were presented. | <p>No RT differences. Accuracy: DD sig lower go% correct, and a lower no-go% correct and overall task performance than TD and FXS (post hoc). No accuracy difference between TD and FXS. Brain Activation- task performance in FXS was associated with left VLPFC, and not by right fronto-striatal region activation. Interactions between FMRP and right striatal dysfunction are associated with this putative compensatory left VLPFC activation in FXS. The extent of right fronto-striatal dysfunction in FXS is correlated with FMRP levels.</p> |
| Tamm et al. (2002) | <p>FXS: $N = 14$, female. IQ WISC-III or WAIS: $M = 84.43$, $SD = 15.79$; TD: $N = 14$, female IQ: $M = 117.93$, $SD = 13.21$; Child Behavior Checklist (CBCL) scores: TD had lower attention and thought problem subscale score. Participants ranged in age from 10–22 ($M = 15.43$, $SD = 3.79$). IQ higher in TD.</p> | CA matched while IQ was controlled. | fMRI | Two kinds of Stimuli conditions: Neutral condition: the word <i>fish</i> was presented 1, 2, 3, or 4 times on the screen (15 trials). Interference condition: Subjects were presented the words <i>one</i> , <i>two</i> , <i>three</i> , and <i>four</i> , presented one, two, three, or four times on the screen (15 trials). TASK (Stroop): Press a button that corresponded to the number of words on the screen. | <p>Behavioral results: FXS more affected by the interference condition (slower RT). Evidence that individuals with FXS sacrificed speed for accuracy, while the controls did not. Brain activation: Both groups recruited the prefrontal cortex (middle and inferior gyri) during the interference condition, for FXS this activation was bilateral, while controls' activation was primarily in the left hemisphere. Between-group comparisons indicated that FXS did not show greater activation than controls in any brain area.</p> |

(continued)

TABLE 1.1 (Continued)

| Citation | Diagnosis and comparison group (<i>N</i> , age, IQ)* | Matching* | EEG or fMRI | Stimuli and task | Findings (RT = Reaction time) |
|-----------------------------|---|----------------------------|-------------|---|--|
| Van der Molen et al. (2012) | FXS: <i>N</i> = 16 males, Age: <i>M</i> = 29.6, Range = 18–42 years; Raven Standard Progressive Matrices RAW SCORES: <i>M</i> = 19.88, <i>SD</i> = 8.15, Raven IQ was equivalent to an average mental age of 7.73 years (SD = 1.59). TD: <i>N</i> = 20 males, Age: <i>M</i> = 29.2, Range = 19–47; Raven Standard Progressive Matrices <i>M</i> = 55.65, <i>SD</i> = 3.76; Raven Standard Progressive Matrices Raw scores were sig. lower in FXS than TDs. | CA matched. CA matched. | EEG EEG | 75 ms 1000 Hz and 1500 Hz sinusoidal tones with 5 ms rise and fall time, presented through headphones, at 80 dB SPL. The interstimulus interval was 1,000 ms. To counterbalance which tone was deviant/standard for each participant, half of the participants were presented with the 1000 Hz tone as standard and the 1500 Hz tone was deviant (and vice versa for the other half). Task: All participants watched a silent movie and were told that the auditory stimuli were irrelevant. | N1: amplitudes to standard tones were larger in FXS males than in controls but larger for deviant tones than standards for the comparison participants. N1 Habituation: In controls, amplitude of the N1 elicited by late standards was significantly smaller than the amplitude elicited by early standards (mean difference = -1.20 P2: amplitudes were significantly larger in FXS than in control group to both standard and deviant stimuli). P2 amplitudes were largest at Cz and smallest at Fz in controls and largest at Cz and Oz in FXS males. |
| Van der Molen et al. (2012) | FXS: <i>N</i> = 16 males, Age: <i>M</i> = 29.6, Range = 18–42 years, Raven Standard Progressive Matrices RAW SCORES: <i>M</i> = 19.9, <i>SD</i> = 8.2. IQ couldn't be calculated but performance was equivalent to an average mental age of 7.7 years (SD = 1.6), PPVT 3rd edition (average verbal mental age of 9.1 years, <i>SD</i> = 2.7. TD: <i>N</i> = 22 males, Age: <i>M</i> = 29.2, Range = 19–47, Raven Standard Progressive Matrices: <i>M</i> = 55.65, <i>SD</i> = 3.8; Raven IQ: <i>M</i> = 121.5, <i>SD</i> = 25.8). Raven Standard Progressive Matrices Raw scores were sig lower in FXS than TD. | CA matched. | EEG | Acoustic stimuli: 1000 Hz and 1500 Hz sinusoidal tones with a duration of 100 ms, including 5 ms rise and fall times. Visual stimuli: blue and yellow colored smiley faces that were viewed from 70 cm distance and presented on a black background in the center of a 17-inch laptop. TASK: A target (deviant) or a nontarget stimulus (standard) was presented for 100 ms (similar in both auditory and visual tasks). Participants were required to respond to the target stimulus (blue or yellow smiley face; 1,000 Hz or 1,500 Hz tone). Responses were registered within the 100–1200 ms response window after stimulus onset. Yellow smiley faces and 1000 Hz tones were used as target stimuli. Participants were instructed to respond as quickly and accurately as possible to the onset of a deviant stimulus by pressing a button on the computer. | Behavioral data: FXS males showed more false alarms, and were slower on both the visual and auditory tasks than controls. FXS males committed significantly more false alarms on the auditory than on the visual task suggesting that selective attention is more compromised in the auditory than in the visual modality in FXS. N1 auditory: N1 amplitude was larger for both stimuli in FXS males compared with controls; no group differences in P2 auditory amplitude were noted. N1 visual: peak amplitudes were maximal at Oz in control participants and at FCz in FXS males. N1 amplitudes were significantly larger in FXS males than in controls for both stimuli, but only at FCz. No differences in Visual P2 were noted between groups. Visual P3b: P3b component was larger in controls than in FXS males, in both auditory and visual modalities. For FXS males, the P3b component was significantly larger in the visual modality as compared with the auditory modality. |
| Castren et al. (2003) | FXS: <i>N</i> = 4 boys, Age: <i>M</i> = 11.6, <i>SD</i> = 2.8, Range = 7–13; and one 21 years old. TD: <i>N</i> = 4 boys, Age: <i>M</i> = 10.6, <i>SD</i> = 0.6 | CA matched. | EEG | (1) Random stimulus sequence of tones consisting of 85% standard (800 Hz) and 15% of deviant (560 Hz) tones. (2) Trains of four identical standard tones. The recordings were performed while the children were watching a silent cartoon on a TV screen, and they did not attend to the tones. | (1) N1 amplitude is larger in children with FXS than in controls. (2) The first half of N2 component for controls is significantly more negative than that of FXS. (3) A significant difference between groups was present in the global-field-power waveforms implying differences in the total brain activity. (4) The topographic distribution of the N1: the N1 peak in children with FXS was well defined when compared with that of controls, maximum amplitude was located frontocentrally. The N1 scalp distribution in FXS patients was similar to controls, whereas the N2 in FXS was located more centrally than controls. |

Down syndrome

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| Wetter et al. (1999) | DS: $N = 20$ Adults, Age: $M = 26.0$, $SD = 10.0$; IQ: $M = 52.5$, $SD = 7.1$; Dementia was assessed using the Dementia Rating Scale-DRS: $M = 99.1$, $SD = 17.9$, $TD = N = 20$, Age: $M = 25.7$, $SD = 10.3$. IQ was not administered to TD group | CA matched; Within group DS assessing relationship between olfactory ERP and dementia. | EEG | Olfactory stimuli (banana odor) were presented via an olfactometer. 2-alternative (odorant and blank), forced choice, ascending method of limits (increased concentration of banana odor) ERP measured at the Fz, Cz, Pz sites for 2000 ms (500 ms prestimulus and 1,500 ms post). | OERP components: the amplitudes and latencies of the OERP components N1, P2, N2, P3, and the interpeak amplitudes N1-N2, N1-P3 were analyzed. Latency: DS demonstrated delays in initial sensory processing. DS demonstrated longer latencies at all components compared with TD. In addition, N2 latency showed a significant effect of age ($F(1,37) = 6.64$, $P < 0.005$), with latencies increasing from adolescence to adulthood. OERP amplitude—not sig. Odor Threshold: DS group had higher thresholds (poorer sensitivity) than TD. Negative correlation between smell threshold and the latency of early OERP components (N1, P2 and N2). Dementia rating scale—negative correlation between DRS scores and P3 latency at all electrodes. |
| Yoder et al. (2006) | DS: $N = 10$ children (6 males). Age: $M = 5.9$, $SD = 1.4$, range 4.33–8.25 years. Nonverbal MA- Leiter-R age equivalency $M = 3.5$, $SD = .8$. Nonverbal IQ: $M = 66.7$, $SD = 8.7$. Grammatical morphology comprehension: $M = 3.7$, $SD = 1.9$; Percentile ranking on Grammatical Morphology subscale of the Test of Auditory Comprehension of Language-TACL-3 ($M = 5\%$, $SD = 4.6\%$) indicated that the sample was quite impaired as a whole ($M = 5\%$, $SD = 4.6\%$), with all but 1 participant falling below the 10th percentile. No control group. | DS split into two subgroups based on their performance on the TACL-3. Then DS was compared based on age, IQ and MA. | EEG | A passive task with three contrasts of consonant-vowel syllables were selected: (1) ga vs. da; (2) ga vs. ba; (3) da vs. na. | ERP difference waves to two consonant-vowel syllables were strongly associated with the degree of impairment in morphological comprehension. Results demonstrated a greater differentiation between event-related potential waves for the less impaired subgroup than the more impaired subgroup within the targeted latency range indicated by the temporal principal components analysis factor of interest. Age, IQ, and MA were nonsignificantly correlated with TACL-3. |
| Lalo et al. (2005) | DS: $N = 20$ Adults (10 males). Age: 18–31, $M = 24.10$ years, $SD = 49$ months. TD: $N = 20$ adults (10 males), Age 19–31; $M = 25$ years, $SD = 44$ months. | CA matched. | EEG | 3 conditions; passive, simple—required to press a button on the armrest chair when detecting target tones and complex — required to aim with their dominant hand from the armrest at a target fixed on their chest when detecting target tones; presented as oddball. | (1) Passive condition: N1 and P2 components were reliably present in all participants. TD: infrequent stimuli usually triggered a MMN under the passive condition. In addition, a P3a component was elicited in about half of the participants, while a P3b was observed in 70%. In comparison, these three components were observed in only 10–25% and their latency tended to be longer in the DS group. (2) Active condition: TD- components N2b and P3b were usually seen under the active conditions, whereas in the DS group the N2b was often missing or delayed. The P3b was present in most participants with DS (55–65% compared with 75–85% in the TD group), albeit with a latency longer than the TD. RT = was shorter in TD than in DS. In addition, RT was significantly longer under the simple > active condition for the DS group only. |

(continued)

TABLE 1.1 (Continued)

| Citation | Diagnosis and comparison group (<i>N</i> , age, IQ)* | Matching* | EEG or fMRI | Stimuli and task | Findings (RT = Reaction time) |
|----------------------|--|---|---|--|--|
| Jacola et al. (2011) | DS: <i>N</i> = 8, 5 female. Age: 18, SD = 3, Range = 12–26. IQ: Stanford-Binet Intelligence Scale, 5th Edition (SB-V). Full scale IQ: <i>M</i> = 44, range = 40–50. Peabody Picture Vocabulary Test (PPVT-III): <i>M</i> = 57, range = 40–83. TD: <i>N</i> = 12, 5 female. Age: <i>M</i> = 19, range = 12–26 years. SB-V full scale IQ, <i>M</i> = 106, range = 88–124. PPVT-III: <i>M</i> = 107, range = 95–133. | CA matched. | fMRI | Experimental condition: press a button for pictures of farm animals. Control condition: Scrambled animal images were presented and participants pressed a button for each image. | Qualitatively and quantitatively different pattern of activation for DS in comparison age-matched TD. Increased activation in DS bilaterally for middle frontal gyrus and regions of the left parietal lobe. An overall reduction in spatial extent of neural activation and atypical patterns of activation were found in individuals with DS. |
| Williams Syndrome | | | | | |
| Haas et al. (2009) | fMRI Study: <i>N</i> = 27, 13 TD, 5 males; 14 WS, 7 males. EEG Study: <i>n</i> = 70, 25 TD, 11 males; 30 WS, 11 males; 15 with developmental delay, 8 males. Both fMRI and EEG: 11 WS participated in both, 4 males. Age: <i>M</i> = 31.13, (SD = 7.55). Williams Syndrome: <i>N</i> = 33, 13 males. Age, <i>M</i> = 31.01 (SD = 8.80). Comparison Group 1 (TD): <i>N</i> = 38, 18 males. Age, <i>M</i> = 29.71, SD = 9.52. IQ: <i>M</i> = 106 (SD = 11.8). Comparison Group 2 (idiopathic ID but not autism): <i>N</i> = 15, 8 male. Age: <i>M</i> = 28.07, SD = 8.8; IQ: <i>M</i> = 62, SD = 8.8). WS and DD were matched on full-scale IQ (WS: <i>M</i> = 65, SD = 6.8, DD: <i>M</i> = 62, SD = 8.8). | CA matched to TD, IQ matched for DD. No significant between groups' ages or proportion of females to males. | Two separate studies, 1 fMRI and one EEG. | Color images of males and females with: happy, fearful, or neutral expressions. Participants identified if each face was male, female, or scrambled by button press. | Behavioral findings: Williams syndrome slower and less accurate compared with age-matched participants but equal to MA matched participants. fMRI and EEG findings: participants with Williams syndrome exhibited heightened amygdala and cortical responses to happy/neutral. WS—exhibited greater right amygdala in happy condition compared with neutral condition. TD—did not exhibit greater left or right amygdala activation to happy condition compared with neutral condition than the WS group. WS exhibited significantly greater right amygdala reactivity to happy compared with neutral. TD: No significant left or right amygdala activation in response to happy compared with neutral was observed. Fearful/neutral group differences: TD exhibited greater right amygdala activation than WS. TD exhibited greater right amygdala reactivity to fearful compared with scrambled faces (nonsocial) at the corrected statistical threshold. There was no significant difference in amygdala reactivity to neutral relative to scrambled faces between the WS and TD groups. <i>Amygdala with IQ, RT and accuracy:</i> (1) IQ as a covariate with BOLD signal extracted from the right amygdala yielded no significant correlations between IQ and amygdala activity either in response to happy ($p = 0.57$) or fearful ($p = 0.48$) facial expressions within the WS group. (2) RT and accuracy as covariates with BOLD signal extracted from the right amygdala yielded no significant correlations |

between either RT or accuracy and activity either in response to happy or fearful facial expressions within the WS group. ERP happy condition:

P300–500-condition \times group interaction indicated that the P300–500 response to happy versus neutral facial expressions was significantly different between the three groups. Happy minus neutral facial expressions was larger for the WS than for the TD group and the DD group (but did not differ between the TD and DD groups). ERP fearful condition: P500–700-condition \times group interaction indicated that the N200 to fearful versus neutral facial expressions was significantly different between groups in mean amplitude. No condition \times group effects for peak latency. Between-group comparisons showed that the N200 difference wave to fearful minus neutral facial expressions tended to be greater (more negative) for the WS than for the TD group over the left hemisphere and was greater (more negative) for WS than for the DD group. In contrast, the effect was smaller (more positive) for the DD than for the TD group. TD group: The N200 did not differ in mean amplitude to fearful versus neutral expressions or peak latency. DD showed increased N200 amplitudes to fearful relative to neutral faces.

(1) Behavioral data: Mean RT: 981.46 ms and accuracy rates: 72.16%. No condition differences (fearful, happy, neutral and scrambled) in RT ($F = 47, P = .70$) or accuracy ($F = .08, P = .97$). In addition, no relations between either RT or accuracy and the sociability measures were found. (2) Indiscriminate sociability and amygdala response to facial expression: Social approach scores were regressed against amygdala response to facial (fearful, happy and neutral combined) expressions compared with scrambled images. No significant positive or negative relationships between social approach scores and either left (group averaged ROI: $R2 = .33, p = .40$; standardized ROI: $R2 = .27, p = .30$) or right (group averaged ROI: $R2 = .11, p = .70$; standardized ROI: $R2 = .04, p = .61$) amygdala response to facial expressions compared with scrambled images, even when age, sex, and handedness were entered as covariates. Furthermore, no significant relationships were observed between social approach scores and either hippocampus and parahippocampal gyrus response to facial expressions.

Colored pictures of headshots of young adults displaying fearful, happy or neutral expressions and scrambled images. Stimuli presented using an event-related design with the four experimental conditions and a resting baseline. Subjects were instructed to judge if each face was either male or female or scrambled by responding with their index finger.

WS: $N = 12$ Adults (8 Female, 4 Male) Age: $M = 29.46, SD = 8.07, Range = 18.03–43.58$

No comparison group.

fMRI

TABLE 1.1 (Continued)

| Citation | Diagnosis and comparison group (N, age, IQ)* | Matching* | EEG or fMRI | Stimuli and task | Findings (RT = Reaction time) |
|------------------------------|---|--|-------------|---|--|
| Thornton-Wells et al. (2011) | WS: N = 10, 6 females. Age: M = 21.8, SD = 1.0 IQ: Kaufmann Brief Intelligence Test-2: M = 79.20, SD = 17.84; 2 control groups of TD: selected cutoff scores (inhibited (IT) ≥ 2.6, uninhibited (UT) ≤ 1.9) Temperament (IT)-N = 10, 6 females. Age: M = 24.3, SD = 1.93 IQ: Kaufmann Brief Intelligence Test-2: M = 117.30, SD = 13.49. 2-Uninhibited Temperament (UT)-N = 10, 6 females. Age: M = 24.5, SD = 1.98; IQ: M = 108.33, SD = 9.03. | TD from each temperament group, based on the traits of IT and UT were matched on gender to WS group. | fMRI | Social and nonsocial images across three emotions (happy, fear and neutral) in a passive viewing task. | WS-IT: Fear nonsocial images: WS > IT controls in left amygdala relative to the IT control group even in relation to neutral expressions. No differences for other comparisons WS (neutral and happy nonsocial images). WS-UT: when viewing social images (faces). Larger left amygdala responses in WS group for neutral social images, compared with the UT control group. When comparing the happy faces to the neutral faces, the UT group had significantly greater left amygdala response than the WS group. |
| Golarai et al. (2010) | WS: N = 13, Age: M = 29.9, SD = 2.6, TD: N = 13, Age: M = 30.0, SD = 2.3. IQ (WAIS-R or WAIS-III) WS: performance IQ: M = 71.08, SD = 2.3, Verbal IQ: M = 64.92, SD = 2.5; FSIQ: M = 66.3, SD = 2.6. TD: performance IQ: M = 117.9, SD = 3.5, Verbal IQ: M = 115.6, SD = 4.6; FSIQ: M = 119.0, SD = 4.2. Benton recognition test on upright face: WS: N = 13; M = 20.8, SD = 0.7. TD: N = 9 of 13; M = 22.63, SD = 1.05. | CA matched and matched Benton face recognition scores. | fMRI | Passive viewing of grayscale images of faces (male, female, various ages, races, expressions and views), objects (abstract sculptures), places (indoor and outdoor), textures (created by randomly scrambling object pictures into 225, 8 x 8, pixel squares). | (1) Average rFFA (right Fusiform Gyrus) volume was two times larger and left FFA volume was 2.5 times larger among WS than TD. The proportional volume of the FFA relative to the anatomical volume of the FUS (fusiform) gray matter was also significantly higher in WS than in TD even when matched on total volume. (2) Variation in the absolute volume of the FFA within each group did not correlate with the anatomical volume of the FFA, gray matter content, or subjects IQ or age. |
| Mimura et al. (2010) | WS: N = 7, all females. Age: M = 34.0, SD = 12.17, All were ELN negative on chromosome 15 by FISH. All exhibited the medical or clinical features of the WS phenotype including cognitive, behavioral, and physical profiles. IQ Wechsler Adult Intelligence Scale third edition (FSIQ: M = 68, SD = 8.7; VIQ: M = 74, SD = 8.7; PIQ: M = 65, SD = 8.8). TD: N = 7, all females. Ages (M = 35.7, SD = 11.1). IQ: FSIQ: M = 115SD = 12.0; VIQ: M = 112, SD = 14.5; PIQ: M = 114, SD = 8.2. | CA matched. | fMRI | (1) Match Affect Paradigm (MAP): consisted of 90 achromatic face stimuli presented in an event-related FMRI paradigm: Two faces were presented that were either congruent for negative or positive affect or incongruent. Participants determined whether the faces matched or not. (2) Control task: Subjects matched gender (male, female) of neutral faces rather than emotions. | (1) Behavioral performance: No difference in accuracy between WS and TD when controlling for FSIQ. (2) Brain Imaging: (a) WS < TD for negative compared with positive valance face stimuli in both right amygdala and right lateral OFC but WS > TD for right medial OFC for negative compared with positive emotional face stimuli. (b) Medial OFC, WS to TD showed significantly reduced activation to negative face stimuli relative to positive stimuli and greater activation to negative stimuli. (c) No correlations between IQ and whole brain activation for either group: significant positive correlation between right lateral OFC activation and IQ for the positive vs. negative valance contrast in the WS group. (d) Control gender matching task showed nonsignificant effects in all these regions and analysis. |

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| Mobbs et al. (2007) | <p>WS: $N = 10$, 8 females. Ages: $M = 31.10$, $SD = 9.7$, Range = 15.5–48.4, IQ: VIQ: $M = 71.6$, $SD = 7.6$; PIQ: $M = 65.6$, $SD = 8.1$; FSIQ: $M = 67.3$, $SD = 7.5$. TD: $N = 8$, 7 females. Ages: $M = 35.2$, $SD = 10.10$, Range = 24.0–54.7. IQ: VIQ: $M = 113.5$, $SD = 14.7$; PIQ: $M = 111.2$, $SD = 8.1$; FSIQ: $M = 114.2$, $SD = 9.8$</p> | CA matched. | fMRI | <p>A nonverbal global processing task was used: (1) 72 experimental stimuli were either a large concentric triangle or square; consisting of smaller triangles or squares; (2) 72 control stimuli consisted of a large, single-lined triangle or square of the same dimensions.</p> | <p>There were significant IQ differences between the groups. WS were less accurate but not slower than TD. No correlation between IQ and global task RT or accuracy. fMRI: between group analysis—greater activation in WS than TD in right MFD, significantly increased activation in WS was seen in hippocampus and parahippocampal gyrus, right lateral geniculate, insula, thalamus, superior temporal gyrus, middle temporal gyrus, orbital frontal cortex, middle frontal gyrus and medial aspects of the precuneus. No significant correlation was found between activation and either IQ or performance accuracy in WS.</p> |
| Key and Dykens (2011) | <p>WS: $N = 21$, 12 males. Age: $M = 26.24$, $SD = 8.26$. IQ: Kaufmann Brief Intelligence Test-2 = $M = 74.25$, $SD = 17.19$; TD: $N = 16$, 7 males. Age: $M = 29.64$, $SD = 11.77$. IQ scores for TD participants assumed to be in the normal range but not tested.</p> | CA matched. | EEG | <p>Oddball paradigm with letter stimuli that could appear as a global form (big letter) or a local element (small letter) but never in same trial. Participants responded yes/no regarding the presence of a target (H) that could occur at either the global (20% probability) or the local level (20% probability).</p> | <p>RT: TD showed global targets associated with shorter RT than standard stimuli or local targets and no RT differences between local targets and standard stimuli. However, WS RT data failed to identify any differences among the conditions. Accuracy rates for TD across all three conditions were near perfect. WS detected global targets more accurately than local targets. ERP: TD: trend for reduced P1 amplitude P1 amplitude for global targets compared with standard stimuli). WS: relative to the standard stimuli, global targets elicited a smaller P1 peak with shorter latency. TD condition differences were present for the centro-parietal P3b amplitude while WS showed no significant effects for the centro-parietal P3b amplitude.</p> |
| Levitin et al. (2003) | <p>WS: $N = 5$, 2 males. Age: $M = 28.8$, $SD = 14.6$; IQ = $M = 63.0$, $SD = 17.2$. TD: no information provided</p> | Age, handedness, gender, and musical experience matched depending on what they measured. | fMRI | <p>Passive listening of noise, music, and silence.</p> | <p>TD: consistent bilateral activation in the superior temporal gyrus and middle temporal gyrus which were not present in WS. WS activations were more widespread and diffuse, recruiting regions in the amygdala and cerebellum. Both groups displayed bilateral temporal lobe activation for music compared with noise and rest. However, these activations were decreased for WS than TD.</p> |
| Paul et al. (2009) | <p>WS: $N = 17$, 10 females. Age: $M = 30.6$ years. IQ: Wechsler: Full Scale IQ $M = 67.5$, $SD = 11.3$ range 47–82. Verbal IQ $M = 72.8$, $SD = 7.8$, range 59–89. Performance IQ: $M = 64.4$, $SD = 10.3$, range 44–83. Peabody Picture Vocabulary Test age equivalents: $M = 12.6$, $SD = 4.5$, range 5.8–22. Visuomotor Integration (VMI) Nonverbal: Figure Drawing Age Equivalent: $M = 5.69$, $SD = 1.6$, range 4.1–11.3. TD (CA matched group): $N = 17$. Age: $M = 31.0$, $SD = 11.2$. TD: MA-matched: $N = 17$, 9 females. Age: $M = 8.8$, $SD = 0.7$.</p> | CA and MA (two different comparison groups). | fMRI | <p>Visual matching of facial identity of black and white photographs appearing in one to 12 possible positions. Two faces sequentially, followed by a delay 500 or 1750 ms delay. A 3rd face appeared and participants indicated whether the test stimulus matched either of the two reference stimuli by hitting the yes or no button.</p> | <p>Behavioral Findings: WS slower and less accurate than CA but not MA comparison participants. Functional neuroimaging: fMRI responses were generally similar in WS participants and MA controls with both groups displaying less robust activations than CA controls. Amygdala response in WS participants showed no activation in contrast to both CA and MA comparison participants.</p> |

the other half involved ERPs or MEGs to assess basic auditory processing. Although there is very little overlap with respect to tasks across studies, both the Stroop task and the go/no-go tasks can be considered measures of executive function (specifically inhibition), while the emotion identification task in females and the eye-gaze direction detection task in males with fragile X can be considered as assessing the social-cognitive correlates of the disorder. The auditory ERP studies allow researchers to assess the integrity of the auditory pathway as well as the ability of individuals with fragile X to habituate to sounds and respond to auditory novelty. These three areas are all considered to be fundamentally impaired in the phenotypes of both males and females with fragile X syndrome (e.g., Hagerman, 1996; Turkstra, Abbeduto, Meulenbroek, 2014).

Auditory Processing Assessed by ERP and fMRI Individuals with fragile X demonstrate sensory difficulties, and specifically auditory hypersensitivity, as measured by parent report. These behavioral findings have been partially substantiated by both MEG (Rojas, Benkers, Rogers, Teale, Reite, & Hagerman, 2001) and ERP studies (for a recent review of electrophysiological findings among individuals with fragile X syndrome in which developmental level is considered in the interpretation of findings, see Knoth & Lippé, 2012). Findings are consistent for certain ERP components which include an overall increased amplitude of the N100, an automatic component elicited by the onset of a sound (Castren, Paakkonen, Tarkka, Ryyanen, & Partanen, 2003; St Clair, Blackwood, Oliver, & Dickens, 1987; Rojas et al., 2001; Van der Molen, Van der Molen, Ridderinkhof, Hamel, Curfs, & Ramakers, 2012, 2012), albeit with findings of decreased habituation of the N100 (Castren et al., 2003; Van der Molen et al., 2012) in relation to chronologically age-matched participants. In addition, a consistent attenuation of the P300, which indexes attention and/or orientation to novelty, has been noted in relation to typically developing CA-matched comparison participants (St Clair et al., 1987; Van der Molen et al., 2012, 2012) but not in relation to MA-matched individuals with Down syndrome (St Clair et al., 1987), indicating that the physiological responses of individuals with fragile X are of similar amplitude to those of individuals of a similar MA. These findings suggest that individuals with fragile X overrespond to sound, and do not habituate (decrease in response to the repeated presentation of a sound) to auditory stimuli in the same manner as typically developing individuals, although the P300 findings of differences relative to CA- but not MA-matched participants

highlight that matching strategies need to be considered in all interpretations and conclusions.

In addition to findings of consistent atypicality in early physiological responses to auditory stimulation, inconsistent findings are noted for other ERP components, including the P2, a physiological response that is greater for infrequent simple targets than frequent targets or frequent distractors, and the N2, which is greater for attended versus unattended stimuli and can index the detection of novelty, stimulus identification, or shifts in attention. St Clair et al. (1987) found increases in P2 but no difference in N2 in relation to typically developing comparison participants using a passive oddball task in which the participants watched a silent movie while two different sounds (one, the frequent, occurring on 80% of the trials and the other, the oddball, occurring on 20% of the trials) were played in the background. In contrast, Van der Molen et al. (2012) found an increased P2 and an increased N2 using a similar passive task. In a subsequent study, Van der Molen et al. (2012) required participants to actively detect oddball targets, and found no P2 differences but an increased N2 in relation to CA-matched typically developing comparison participants. These findings suggest that processing differences depend on whether participants are actively engaged in an experiment or passively listening, which may or may not be related to MA. The implications of these studies are diminished by small groups and the use of CA matching (with the exception of St Clair et al., 1987, who compared individuals with fragile X to both CA-matched typically developing peers as well as MA-matched peers with Down syndrome).

In the only MEG study conducted with participants with fragile X, the participants watched a silent movie while pure tones were played, thereby allowing for the assessment of basic auditory processing (Rojas et al., 2001). (Briefly, MEG is a technique that combines some of the temporal resolution of ERPs with the some of the spatial resolution of fMRI.) Rojas et al. compared 11 participants with fragile X (six female, five male) to typically developing peers matched on CA and reported an increased amplitude of the mN100 component, the MEG analogue of the auditory N1 in EEG studies, among the participants with fragile X, suggesting greater neuronal activation in response to sounds. However, these findings need to be qualified due to the differences in developmental level between the groups, as the IQs of the typically developing comparison participants were more than double that of the participants with fragile X, and well above the average range. Given these vast differences in IQ between the groups and the notion that CA affects the amplitude of ERP responses (Gomes et al., 2001), we need

to consider whether the amplitude differences between the groups might reflect meaningful phenotypic differences or are a confound arising from the significant differences in developmental level of functioning (i.e., MA) between the groups. Also given the IQ differences between males and females with fragile X, whose ERPs were averaged together into one group, gender differences may have confounded the findings. If MA affects the magnitude of ERPs, then averaging females with high MA (and their characteristic ERPs) with males with low MA (and their characteristic ERPs) may have led to an average ERP response that is characteristic of neither the males nor the females in the sample. Accordingly, the group-level analyses comparing typically developing and fragile X groups may not reflect any meaningful differences.

In spite of these major caveats, other evidence has also supported the notion that auditory processing in fragile X is atypical. For example, Castrèn, Paakkonen, Tarkka, Ryyanen, and Partanen (2003) found higher N100 amplitudes using a similar passive paradigm and ERPs among a small sample of children with fragile X compared with CA-matched typically developing children, providing further support for the notion that basic auditory processing appears to be atypical among individuals with fragile X. However, these findings must be considered in light of the difference in level of functioning between the participants with fragile X and the typically developing participants and very small group sizes. Nonetheless, ERP and MEG findings of basic auditory processing in fragile X are consistent with those of the fMRI knockout mouse, an animal model of the disorder, which also suggest auditory hypersensitivity. In addition, the knockout mouse also shows deficits in cerebellar learning that are temporally sensitive, a key factor in auditory processing (Chen & Toth, 2001).

Based on these links between ERP findings of basic auditory differences and mouse models of fragile X, Hall, Walter, Sherman, Hoefl, and Reiss (2009) sought to assess the ability of females with fragile X and MA-matched typically developing female participants to make auditory temporal discriminations and measured the relationship between this ability and activations in sensory areas using fMRI. Although no differences in behavioral performance as measured by response time and accuracy were noted between the participant groups in their ability to assess whether a comparison tone was longer or shorter than a standard tone, fMRI activations differed between the groups.

The activation differences were compared by subtracting activations to the task condition described above with

a control condition in which the participants were asked to respond to a button press after the presentation of two identical tones. Hall et al. (2009) noted that these two conditions were identical in every way, except that in the control condition the participants did not have to make temporal discriminations (the tones were always of equal duration, though participants were not told this). The primary finding was that subtracting activations between the discrimination and control tasks yielded more diffuse areas of activation in frontal, temporal and limbic regions for the participants with fragile X, whereas for the typically developing participants, the subtraction led to focal activations in parietal and occipital regions. This latter finding is somewhat surprising as Hall et al. were trying to isolate an auditory temporal discrimination but report activations in both the parietal lobe which is related to tactile processing, number knowledge, and multisensory integrations and in the occipital lobe which houses the visual regions, neither of which should have been involved in the task processing. Direct group comparisons yielded increased left hemisphere activations for the participants with fragile X in the frontal (left medial frontal gyrus) and temporal (left superior and middle temporal gyrus, left cerebellum and left pons) regions. No areas of the brain were more active among the typically developing than among the participants with fragile X, suggesting that the participants with fragile X used more processing resources than the typically developing participants. Hall et al. examined the associations among IQ, age, and activation levels and noted that their findings could not be attributed to these potentially confounding variables because the correlations among age, IQ, and activated cortical and subcortical regions were not significant.

Hall et al. (2009) also found that task performance correlated with cerebellar activations in the typically developing group but with middle temporal gyrus for the fragile X women, and that contrast values of the significantly activated brain regions did not correlate with levels of fMRP among the females with fragile X. These findings suggest differences in activation patterns on this simple task despite similar behavioral performance, and that these differences were related neither to IQ, age, nor the severity of fragile X, as genetically defined. However, the lack of significant correlations could also be due to a restricted range in these variables, the small sample size, or the lack of correction for multiple comparisons in their fMRI results, a problem which plagues fMRI research and which some authors suggest will always lead to increases in false positive discovery rates and subsequent difficulties in interpreting findings (see Bennett, Wolford, & Miller, 2009; Poldrack, 2012).

Overall, simple temporal discrimination abilities among females with fragile X syndrome appear commensurate with MA, although task performance seems supported by different brain regions, which could reflect either increased mental effort or some sort of compensatory processing.

Gaze Processing as Assessed by fMRI Garrett, Menon, MacKenzie and Reiss (2004) and Watson, Hoefft, Garrett, Hall and Reiss (2008) conducted similar studies of gaze processing among females and males with fragile X, respectively. Gaze avoidance is a hallmark symptom of fragile X even in the absence of a comorbid diagnosis of ASD, and generally occurs across both males and females with the disorder. Both studies involved similar stimuli, task, and methods in which participants were presented with pictures of faces (angled or forward facing) whose gaze was either direct or averted. The participants responded by button press as to whether the face was looking at them or away from them. Garrett et al. compared females with fragile X with IQs in the average range to typically developing females matched on age. However, despite the attempt to include only females with fragile X with average IQs, the IQs of the typically developing comparison participants were significantly higher than those of the fragile X participants. In a different approach, Watson et al. compared boys with fragile X to both typically developing children of the same CA and individuals with a developmental delay (DD; group composition not mentioned but specified as not autistic) matched on the basis of both age and IQ. With respect to task performance (RT and accuracy), the typically developing groups in both studies performed better than the individuals with fragile X. The boys did not differ in performance from the IQ matched group of DD participants, but the importance of this finding is negligible due to the problems in methodology and interpretation associated with the use of mixed etiological comparison groups. A significant correlation between IQ and task performance was noted for females, but not for males when compared with participants of equivalent MAs. That is, both males and females with fragile X appear to display a relationship between performance and developmental level. In neither study were RT differences found across the groups.

Group differences in the processing of direct versus averted gaze between individuals with fragile X and their respective comparison participants were noted in both the Garrett et al. (2004) and Watson et al. (2008) studies. Activations were similar between the participants with developmental delays and the typically developing participants, but activation patterns differed across studies with the exception of the superior temporal sulcus (STS),

which showed greater activation in the typically developing groups as compared with the fragile X boys (Watson, 2008) and girls (Garrett, 2004). The finding of completely different activation patterns, even among the typically developing participants, is puzzling as essentially identical tasks yielded very different findings. The most we can conclude from these fMRI findings is that individuals with fragile X showed a relatively consistent decrease in activation in the STS, an area generally associated with the perception of other's gaze (Campbell, Heywood, Cowey, Regard, & Landis, 1990) and biological motion (Grossman & Blake, 2002).

Overall, the findings from the neuroscience studies of individuals with fragile X suggest both that auditory processing appears atypical in relation to CA-matched participants but not in relation to MA-matched participants (at least for the P300) and that gaze processing appears atypical in fragile X in relation to both CA- and MA-matched comparison participants. MA among persons with fragile X was, as expected, consistently linked to behavioral performance, but less so in relation to brain activations related to behavioral tasks.

Persons with Down Syndrome.

Simple Sensory Processing in Down Syndrome Using ERPs Many ERP studies of participants with Down syndrome have involved no-task paradigms in which participants just passively listen to sounds or view pictures while brain activity is recorded. This allows for the assessment of basic sensory function and is generally used to assess afferent (bottom-up) rather than efferent (top-down) pathways without requiring participants to complete an experimental task. The advantage of these studies is that they can be conducted in lower functioning individuals and are not tied to cognitive functioning. The disadvantage, of course, is that they are not informative about cognitive, or any other aspect of, functioning. Briefly, evidence from passive studies of auditory and visual functioning suggests that basic auditory processing of tones among individuals with Down syndrome is atypical, with reports of longer N1 latencies and higher P2 amplitudes in infants relative to CA-matched typically developing infants (Barnett & Lodge, 1967; Seidl, Hauser, Bernert, Marx, Freilinger, & Lubec, 1997), as well as an attenuated habituation to visual stimuli in frontal regions (but habituation was observed in other regions as well as behaviorally; Karrer, Karrer, Bloom Chaney, & Davis, 1998; Karrer, Wojtascek, & Davis, 1995). These findings appear to be consistent across childhood with findings of an attenuated P3 response during repeated auditory stimulation.

In addition to the studies involving responses to simple tones, Yoder, Camarata, Camarata, and Williams (2006) assessed the relationship between standardized measures of grammatical morphology and ERP responses to different consonant-vowel syllable pairs among individuals with Down syndrome. They divided a group of participants with Down syndrome into high and low grammatical morphology skill sub-groups (although the majority of participants fell below the 10th percentile) and found increased differentiation between syllables in the ERP waveforms among participants in the less-impaired group than among those who were more impaired, suggesting a relationship between grammatical abilities and brain responses. These findings suggest that increased mental effort is required for individuals with Down syndrome in the processing of basic sounds relative to their same-aged peers; that this finding is sustained over age, and that grammatical skills can reliably be differentiated among persons with Down syndrome using ERP technology. No studies of passive processing involved MA matching strategies and so, again, the meaningfulness of these findings are difficult to evaluate.

To assess whether the need to prepare a motor response influences the speed of perceptual auditory processing among individuals with Down syndrome, Lalo, Vercueil, Bougerol, Jouk, and Debu (2005) examined auditory processing and its relationship to movement complexity among adults with Down syndrome and CA-matched typically developing comparison participants. They used passive, simple active (button press responses) and complex active (motor movement to a spot on their chest) versions of an auditory oddball task. Lalo et al. noted RT differences between the groups as the responses under the simple task conditions were longer than the complex task condition among the participants with Down syndrome, but did not differ among the typically developing participants. Auditory N1 latencies were longer and auditory P2 responses were larger for the participants with Down syndrome than for the comparison participants when comparing simple versus complex tasks (with the passive task serving as the baseline). Inconsistent with Lalo et al.'s hypotheses, motor complexity did not appear to affect the ERPs for either group of participants. In addition, the identifiable ERP peaks were noted less reliably overall (for peaks such as the MMN, N2b, and P3b) for the adults with Down syndrome in relation to the typically developing participants, suggesting that the reliability of even simple sensory processing peaks might fundamentally differ between persons with Down syndrome and typically developing people of the same chronological age. The lack of reliable ERP peaks in this population suggests

either a fundamental alteration of processing or could be related to the young mental ages of the participants, as the MMN continues to develop through middle childhood and adolescence (Bishop, Hardiman, & Barry, 2011), as do auditory ERPs more generally (Bishop, Anderson, Reid, & Fox, 2011). Furthermore, the lateralization of ERP components also changes with age, with auditory ERPs become more visible in central scalp locations with increasing age. Since Bishop et al. only recorded ERPs at midline central locations (Cz and Pz), their findings may be related to differences in the location of ERPs among individuals with Down syndrome whose cognitive abilities are less mature than among typically developing participants and whose ERPs would perhaps be larger at lateral electrodes than at central scalp location.

In the one study of olfactory ERPs among adults with Down syndrome in relation to CA-matched comparison participants, Wetter and Murphy (1999) found that individuals with Down syndrome had lower olfactory thresholds (poorer sensitivity to smell), as well as longer latencies to process smell in relation to typically developing participants. For both groups, detection threshold and ERP latency were correlated, and specifically in the group of participants with Down syndrome, higher ratings of dementia were related to longer latencies of the P3b component, suggesting that individuals with Down syndrome who had poorer cognitive functioning had a slower brain response to smell. Overall, sensory processing (visual, auditory and olfactory) appears to be more effortful for individuals with Down syndrome than CA-matched typically developing comparison participants.

Higher Order Processing Among Persons With Down Syndrome Despite the numerous studies of resting state fMRI, which provide information about brain anatomy and the brain's functional organization in the absence of task demands, we only found one fMRI study in which participants actively listened to speech and one in which individuals with Down syndrome completed some sort of experimental task that relates to cognition. Losin, Rivera, O'Hare, Sowell, and Pinter (2009) assessed brain activation patterns using fMRI while participants with Down syndrome and age-matched typically developing adolescent participants rested (control condition), or passively listened to speech presented forward and backward. The contrast of interest was between forward and backward speech, which would allow researchers to isolate the language-specific aspects of receptive language. The main finding was a significant difference in brain activations between forward and backward speech among the typically developing participants (temporal

language regions), but not for the participants with Down syndrome, suggesting that the latter group did not process the linguistic elements of the task.

In contrast to the passive listening approach of the previous study, Jacola et al. (2011) compared brain activations of adolescents and adults with Down syndrome and CA-matched typically developing participants while the participants completed a basic categorization task in which they were presented with pictures of animals one at a time and were asked to respond by button press to farm animals. Jacola et al. compared activations elicited by the categorization task to a control condition in which the participants pressed a button in response to the appearance of a scrambled image (simple detection) to isolate visual object recognition and semantic classification regions of the brain. In addition to the experimental task, the participants also completed the Peabody Picture Vocabulary Test (PPVT; a standardized measure of receptive vocabulary) as well as the Stanford-Binet V (SBV), a measure of IQ. The SBV scores reported reflect only scores on a composite of verbal and nonverbal visual-spatial subtests, which were correlated to the BOLD responses. Correlational analyses between the BOLD responses and the PPVT scores were not computed because of “more variability in the scores for the individuals with Down syndrome compared to those for the typically developing individuals” (Jacola et al., 2011, p. 349). Overall, the mean IQ of the typically developing participants was double that of the participants with Down syndrome, and, as might be expected, the performance of the participants with Down syndrome was much worse than that of the typically developing participants. However, these differences in performance were not correlated with PPVT or SBV scores suggesting that task performance was not related to underlying cognitive abilities. On the basis of these findings, Jacola et al. suggest that individuals with Down syndrome activated different brain areas than the typically developing participants to complete the task, that the relationships between SBV scores and areas of activation differed between the groups, and that activation differences could also reflect performance differences. The findings from this study are difficult to interpret because of small sample size, differences in cognitive abilities between the groups, as well as differences in thresholding requirements for inclusion of BOLD activations between the two groups of participants—for no specified reason, the inclusion practices were more liberal for the participants with Down syndrome than for the typically developing participants. Although a first step in the quest to understand differences in brain activations involving a task that requires cognitive

processing, the findings from this study do not really provide any information other than to say that individuals with Down syndrome are different than CA-matched typically developing individuals.

This review highlights that we are only at the beginning stages of utilizing neuroscience approaches to complement the study of cognitive processing among persons with Down syndrome. Very few studies involved placing any cognitive demands on the participants, and none involved MA-matching. This is particularly troubling as Down syndrome is one of the most common etiologies of intellectual disability, with one of the better-understood cognitive phenotypes. In addition, the findings from all of these studies suggest that the defect approach permeates the literature on Down syndrome. As such, activations in the case of fMRI and neurophysiological responses in the case of ERPs are always considered defective, if they differ from that of typically developing participants who generally have double their IQs. Only Jacola et al. (2011) even acknowledged that MA matching might help further our understanding of how and whether individuals with Down syndrome process information in different ways than typically developing individuals.

Persons With Williams Syndrome. As persons with Williams syndrome are characterized by a hypersocial phenotype and common visuospatial deficits that appear related to a difficulty integrating parts into wholes, neuro-based studies of individuals with Williams syndrome have been focused on global-local stimulus processing ($n = 2$) or amygdala responses to different facial expressions ($n = 6$). Five of the studies reported here were completed by the same research group, headed by Ursula Bellugi and Allan Reiss, who have provided an extensive understanding of both Williams syndrome and of the role of the amygdala more generally. Their group seemed to consistently consider issues related to matching participants, either by assessing the effect of IQ on task performance and regions of interest, by directly matching participants on the basis of MA, or at the very least by acknowledging a lack of a comparison group as a limitation.

Global-local Processing in Williams Syndrome Both studies of global-local processing in Williams syndrome involved the presentation of Navon-type stimuli (Navon, 1977), composed simultaneously of global and local levels. The global level stimuli, typically letters or shapes, are made up of smaller, local level letters or shapes, which could either be congruent or incongruent with the global level (e.g., a large S made up of small s's or small o's respectively). Typically developing individuals generally show a

global precedent on these tasks, as they are faster and more accurate at detecting stimuli at the global level as compared with the local level, although this may vary as a function of stimulus characteristics (e.g., Enns & Kingstone, 1995; Kimchi et al., 2005). Mobbs et al. (2007) used fMRI to assess global–local processing of shapes among participants with Williams syndrome and CA-matched typically developing participants. The participants were asked to attend only to the big shapes and to press one button if the big shape was a triangle and a second button if the big shape was a square. Thus, in this case, the local level of the stimulus was never mentioned to the participants. Comparisons were made between BOLD and performance responses on this experimental task with a control task in which shapes were presented without local elements (e.g., a big triangle and a big square). Mobbs et al. found that the participants with Williams syndrome were less accurate and marginally slower than the typically developing participants, but these results did not seem to be linked to IQ. Activation differences were diffuse, with areas of larger activation among the participants with Williams syndrome in the dorsolateral prefrontal cortex, which Mobbs et al. linked to increased effort required to perform the task. Less activation was seen in parietal and early visual cortical areas among the individuals with Williams syndrome, suggesting disruptions in the dorsal-stream pathway.

Key and Dykens (2011) compared the performance of participants with Williams syndrome with typically developing participants matched on the basis of CA. The participants were asked to respond yes or no (by button press) with respect to the presence of the letter H, which could appear either at the local level (an S made up of small h's) or the global level (an H made up of small s's) with equal probability (20% each) during concurrent ERP recordings. The remaining 60% of trials were non-target standards (which did not contain the letter H). The presentation of only 30 target trials (30 global and 30 local stimuli) is problematic as it is a very low number of trials for an ERP study, which requires multiple repetitions to reduce the signal to noise ratio. Four components of interest were tested for between and within group effects. Early components included the occipital P1 (between 70 and 150 ms) and the N150 (between 150 and 220 ms), whereas later components of interest included the centro-parietal P3b (between 300–600ms) and the frontal P3a (between 200 and 400 ms) based on other ERP studies of global-local processing.

As expected, the typically developing participants demonstrated a global precedent behaviorally as evidenced by faster responses to the global stimuli than both the local and the distractor stimuli, but were equally accurate

across stimulus types. ERP components were significantly different among the standard, local, and global level stimuli at the N150, and P3a and b components. Global and local targets elicited more negative N150s than standards, and more positive P3a and b amplitudes. No differences were noted between the physiological responses to global and local targets. Among the participants with Williams syndrome, no global precedent was noted although the global targets were detected more accurately than the local targets (75% vs. 31% approximately). ERP differences were noted for the early N150 and P1 components. P1 amplitudes were lower for the global versus the standard targets and had a shorter latency. The N150 component was more negative (larger) for the global versus the standard stimuli. Further, later in the processing stream, the global targets elicited a larger and earlier P3a component relative to standard stimuli. Although these findings suggest that individuals with Williams syndrome detect the global configuration of Navon stimuli similarly to typically developing participants early in the processing chain, additional analyses would have allowed for stronger conclusions. One, the groups were never compared with one another, so group differences or interactions between the conditions and group were never assessed. Two, the processing of local targets was not compared directly with the processing of global targets, which would have also allowed for an understanding of the relative physiology underlying global–local processing. Three, the participants with Williams syndrome were compared with college students who, even if their IQs were not assessed, can be assumed to function at a significantly higher cognitive level. Key and Dykens made no attempt to correlate task performance with IQ as a function of condition, nor did they take into account the obvious IQ differences between the groups in their analyses of ERPs or in their discussion (other than to say that IQ matching might be a useful future direction). Although this study provides evidence that early visual processing of global configurations seems typical in Williams syndrome, this study, as with the one by Mobbs et al., does not allow for an examination of the relations between global and local processing. Without actual comparison of these levels of processing, much remains to be learned about the nature of global-local processing among persons with Williams syndrome.

Facial Expression Processing and Face Processing in Williams Syndrome To better understand the hypersociality in this group, Haas et al. (2010) used fMRI to assess the relationship between amygdala responses to different facial expressions (happy, fearful, neutral and control images that were scrambled) among adults with Williams

syndrome. The participants' parents completed a questionnaire used to assess their adult child's sociability across three scales (global sociability, approach strangers, and approach familiars). In the experimental task, the participants who were in the fMRI portion of the experiment were asked to respond by a button press as to whether the image they were presented was male, female, or scrambled (three buttons). The goals were to look at the associations among task performance, BOLD responses in the amygdala, and parental reports of sociability. Behavioral findings indicated no relationship between task performance (reaction time and accuracy) and the parental reports of sociability. However, consistent with the notion that the increased social approach of individuals with Williams syndrome is related to a decreased response to social fear, Haas et al. found that a diminished amygdala response to fearful faces was associated with a greater tendency (as reported by parents) to approach strangers. This is initial evidence of a neural correlate of behavioral observations of hypersociability, and provides a developmental pathway that may lead to this behavioral phenotype.

In a further attempt to understand the relationship between hypersociability and amygdala responses, Paul, Snyder, Haist, Raichle, Bellugi, and Stiles (2009) compared the performance and associated BOLD responses of individuals with Williams syndrome as well as a group of typically developing participants matched on CA and a group of younger typically developing children matched to the participants with Williams syndrome on the basis of MA on a facial expression matching task. The participants completed an experimental task in which they were required to determine whether a target face matched either of two faces presented just prior to the target. This task was completed both in and out of the scanner with slight modifications. Behaviorally, prior to the scanning, the performance of the individuals with Williams syndrome was as fast and as accurate as the MA-matched typically developing children, and both of these groups were slower and less accurate on the task than the CA-matched typically developing participants. However, while performing the task in the scanner, the MA- and CA-matched participants outperformed those with Williams syndrome with respect to both reaction time and accuracy. With respect to BOLD responses, the physiological responses to the task were in many ways similar between the participants with Williams syndrome and the MA-matched group, whose activation patterns were less robust than the CA-matched participants. The exception to this was the amygdala, which was more active in both the MA and CA groups than among the participants with Williams syndrome.

The finding of an underactive amygdala is consistent with Haas et al.'s (2010) finding that decreased amygdala responses to fearful faces are associated with a greater tendency to approach strangers.

Haas, Mills, Yam, Hoeft, Bellugi, and Reiss (2009) used both ERPs and fMRI to test the neural basis of social responsivity among individuals with Williams syndrome in response to emotionally valent facial expressions (happy and fearful). In both the ERP and the fMRI studies, the participants were asked to detect whether the images that were presented were male, female or scrambled by pressing one of three response buttons. The face stimuli depicted happy, fearful or neutral facial expressions. Fourteen participants with Williams syndrome completed the fMRI study and their performance was compared with that of typically developing individuals matched on the basis of CA, while 30 participants with Williams syndrome completed the ERP study (11 overlapped) and their performance was compared with typically developing participants matched on CA and to a smaller group of participants with idiopathic developmental delay (participants who had an intellectual disability of unknown origin and were not autistic) matched on the basis of full scale IQ (test unspecified). The age range of the IQ-matched participants groups were similar, though the authors did not mention that they specifically match on CA. Behaviorally, the performance of the participants with Williams syndrome was slower and less accurate than the CA-matched typically developing participants but did not differ from the IQ-matched participants with idiopathic developmental delay. The fMRI findings indicated that compared with CA-matched participants, the participants with Williams syndrome showed increased right amygdala activation for happy as compared with neutral facial expressions, and lower right amygdala activation than the typically developing participants when the fearful facial expressions were compared with the scrambled images. Neither IQ, reaction time, nor accuracy accounted for their findings.

With regard to the ERP paradigm, Haas et al., (2009) compared differences between happy and neutral facial expression as well as fearful-neutral facial expressions in three time periods (N200, P300–500 and P500–700). At 200 ms, they found that the participants with Williams syndrome showed a larger difference between fearful and neutral facial expressions than the MA-matched participants and a trend for the same finding among the CA-matched participants (but only the participants with Williams syndrome showed a smaller response to fearful facial expressions relative to neutral ones). Between 300 and 500 ms, the participants with Williams syndrome

demonstrated greater physiological responses (larger differences between happy and neutral expressions) relative to both the CA- and MA-matched groups (which did not differ from each other). Between 500 and 700 ms, there was a larger difference between physiological responses to fearful—neutral facial expressions in the CA-matched participants relative to the participants with Williams syndrome. The Williams syndrome and MA-matched groups did not differ in their responses at this time point. Together, these findings corroborate the notion that individuals with Williams syndrome show a decreased response to fearful facial expressions, which is visible early in the processing stream and which appears to be maintained in more cognitive time frames (between 500 and 700 ms). The participants with Williams syndrome also showed increased reactivity to happy facial expressions as measured by both fMRI and ERPs. These findings map well onto the behavioral phenotype of individuals with Williams syndrome who are often very socially driven but also approach others indiscriminately, although the significance of these findings are diminished considerably due to the use of two problematic groups—CA-matched typically developing persons and an etiologically unspecified group of persons with intellectual disability.

Another explanation of the unique social characteristics of persons with Williams syndrome is linked to the finding that they rate pictures of faces as more approachable than MA-matched comparison participants (Frigerio et al., 2006), a phenomenon that Bellugi, Adolphs, Cassady, and Chiles (1999) termed a positive attribution bias. In an attempt to understand this positive attribution bias, Mimura et al. (2010) assessed activations of the amygdala and the orbitofrontal cortex (OFC), which are involved in facial affect recognition among seven participants with Williams syndrome and seven typically developing participants matched on CA. The participants completed a facial expression-matching task by responding with a button press as to whether two faces presented side by side had matching or mismatching facial expressions while in the scanner. Specifically, Mimura et al. attempted to assess the role of both the medial and lateral portions of the OFC, which play different roles in facial expression recognition. The medial regions of the OFC are involved in monitoring the reward value of reinforcers, whereas the lateral OFC is involved in the monitoring of punishers. Mimura et al. hypothesized that among typically developing individuals, positive facial expressions would activate the medial OFC while negative facial expressions would activate the lateral OFC. In contrast, they expected that among individuals with Williams syndrome, facial expressions depicting both

positive and negative facial expressions would activate medial regions as a result of their positive attribution bias. Accuracy on the task did not differ between the groups despite IQ differences (although *p*-values increased when FSIQ was used as a regressor).

Mimura et al.'s (2010) fMRI findings indicated that the participants with Williams syndrome had decreased activation in the right amygdala and right lateral OFC relative to the typically developing participants for negative facial expressions but the right medial OFC showed greater activation than the typically developing participants in response to negative stimuli (compared with positive stimuli). Although no relationship between task performance and IQ were found at the level of the whole brain, when region of interest analyses were repeated in the participants' native space, a correlation was found between right lateral OFC activation and IQ for the positive versus negative contrast among the individuals with Williams syndrome, suggesting that those individuals with higher IQs also had larger activation differences between positive and negative facial expressions in this region than those with lower IQs. These findings are consistent with previous evidence of decreased amygdala responses to negative facial expressions among persons with Williams syndrome as compared with typically developing individuals, but are novel with regard to the finding that that negative facial expressions activate regions of the brain associated with reward gating in individuals with Williams syndrome compared with typically developing participants. This suggests that these two regions might work in tandem to explain why individuals with Williams syndrome tend to approach strangers. That is, individuals with Williams syndrome view negative facial expressions as rewarding, and also show a decreased fear response to negative facial expressions.

Thronton-Wells, Avery, and Blackford (2011) also attempted to dissect the role of the amygdala among individuals with Williams syndrome by comparing BOLD responses to social and non-social images among participants with Williams syndrome relative to comparison participants with an inhibited temperament group (who have high non-social fear responses, which is consistent with the Williams syndrome phenotype) and those with an uninhibited temperament who are characterized (similarly to individuals with Williams syndrome) as being highly sociable. Thus, they attempted to determine whether the typical findings related to amygdala responses among persons with Williams syndrome are unique and specific to this group or rather more characteristic of the personality traits associated with the phenotype. Their findings indicated that individuals with Williams syndrome had

larger BOLD responses to non-social fear images than the typically developing individuals with a similar level of fear. With respect to social images, the BOLD response in the amygdala of individuals with Williams syndrome was commensurate with that of typically developing individuals who are highly social, suggesting that high sociability personality traits might explain more generally the heightened amygdala responses rather than being unique to individuals with Williams syndrome.

In addition to the role of the amygdala and the OFC in facial expression processing, and thus in the hypersociability of individuals with Williams syndrome, the fusiform face area is also an important region associated with the identification and recognition of faces. As individuals with Williams syndrome have been found to have face-identity recognition abilities that are commensurate with CA and better than would be expected for MA (Bellugi, Lichtenberger, Jones, Lai, & St George, 2000), the fusiform face (FFA) area might be involved in Williams syndrome. In one example, Golarai et al. (2010) systematically deconstructed the role of the FFA (and the relationship between task performance, IQ and FFA activation) among individuals with Williams syndrome and CA-matched typically developing comparison participants on a passive viewing task in which the participants were presented with grayscale images of faces, objects, textures and places. They sought alternative explanations to their finding that the FFA of individuals with Williams syndrome is almost double the volume of the FFA of typically developing participants by using different thresholds, clustering and spatial smoothing techniques, and comparing their FFA results to other brain areas associated with face processing. The participants also completed a face recognition task outside of the scanner and, consistent with previous findings, performance on the task did not differ between typically developing participants and those with Williams syndrome, although a significant correlation between size of the right FFA and task performance was found among the participants with Williams syndrome but not among the typically developing participants. This correlation remained significant even after IQ was controlled (despite task performance being correlated with IQ), thereby providing a neural anatomical correlate that might explain why individuals with Williams syndrome have better facial recognition abilities than would be expected for their MA.

Summarizing Neuroscience Work Across Etiologies. Our review of neuroscience-based studies across etiological groups highlights that we are in the most nascent stages of work in this area. The studies were generally characterized

by essential methodological problems either in their execution, or in their consideration of cognitive level in their findings, thereby considerably diminishing the conclusions that can be made.

Although the study of intellectual disability within the context of neuroscience is still a new area of inquiry, several foundational questions have been raised, especially with regard to the roles of developmental level, CA, and specific etiology on ERP and fMRI findings. We have no methodological consensus regarding how best to treat IQ, or cognitive development more generally, nor do we have a real understanding of their relation to brain physiology. Further, the difficulties in recruitment and retention of participants with an intellectual disability for studies involving fMRI and ERPs necessitate that the most of the studies involve adult participants, which represents the endpoint of development, leaving open an important chicken-egg problem. That is, what is the degree to which neural differences are causes or consequences of the behavioral phenotype? Nonetheless, as more researchers with different areas of expertise work together, we hope that these questions as well as those regarding the relationship between brain and behavior in intellectual disability will be addressed.

The current advancement of technology and the related increased access and funding available to researchers using neuroscience tools to study persons with intellectual disability are certainly welcome contributions to this field, but have given rise to a new generation of defect theorists who, in their frantic pursuit to identify key neurological problems, lay waste to many of the tenets of developmental theory and methodology. Thus, decades after its apparent demise, the defect approach appears to be rearing its head again. We are a long way from being able to consider the whole person from a neuroscience perspective but we hope that by promoting the importance of developmental theory and methodology that we will accelerate the developmental legitimacy of neuroscience research in the study of intellectual disability.

CONCLUSIONS

From Genes to Brain to Behavior in Intellectual Disability: Future Directions in Research

More than 10% of the human genome has been implicated in intellectual disability, both organic and familial (Schuurs-Hoeijmakers et al., 2013). The number of well-known syndromes associated with intellectual disability is quite small, however a major advancement of the

twenty-first century is the mapping of these known genetic disorders to neural consequences. This approach has led to breakthroughs in understanding exactly how specific genes work together to build a brain. The burgeoning field of epigenetics is also beginning to shed light on the subtle influences of external factors, from diet and the prenatal environment, to parenting. Each child enters their individual world, with their unique epigenome, acting upon and eliciting experiences from the world that combine to carve the child's unique developmental path, like water carving a stream in the sand. Just as every child carves out a unique developmental pathway, some are more functionally adaptive than others. Thus, the orthogenetic principles and similar-sequence/similar-structure hypotheses are still relevant to understanding both familial and organic intellectual disability, as are Piagetian constructivism, and the neo-Piagetian neuroconstructivist (Karmiloff-Smith, 1998) models in which the child, typically developing or otherwise, is the unique product of cumulative and self-driven experiences. Via general and universal developmental principles, the unique child constructs his or her own path through the selection of experiences and evocations from the environment. The biology of the child presents merely a set, albeit often a powerful one, of constraints.

Summary

Throughout this chapter, we highlight, that, as with all developmental processes (Werner, 1957), the development of the developmental approach to intellectual disability led to an essential deconstruction, in this case of the commonly applied theories and methodologies in the field, as part of the progression toward a more precise but comprehensive understanding. Thus, this deconstruction is not one of a Humpty Dumpty who can never again be put together. Rather it is one of a chrysalis in which the ideas are even more beautiful and lofty than the original. This optimism is based in the advances in the almost half century since Zigler's original articulation of the developmental approach to intellectual disability and in the twenty-first century sophistication in 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 complex developmental trajectories and relations among them. Through this synergy, the key to understanding the development and functioning of the heterogeneous grouping of persons labeled as intellectually disabled is with the paradoxical top-down imposition of developmental theory on the bottom up process of

piecing together a mosaic based on fine-tuned and precise empirical evidence from relatively precise homogeneous groupings. Despite, and because of, the abandonment of the big stories of the defect and related approaches to intellectual disability, we now know much more about persons with intellectual disability 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. As we (Burack et al., 2012) noted elsewhere, the outcome of this process might best described by yet two more paradoxes, that the more we know, the less we know, and that is still progress, because, the less we know we know, the more we (really) know. Accordingly, despite all the advances in research on persons with intellectual disability since the Hodapp and Burack (2006) chapter in the last edition of this handbook, we can make the same claim that we are only in the early stages of the development of the developmental approach to intellectual disability and are still just becoming more aware of the immensity of the task ahead of us—but that is the nature of a developmental process.

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