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Children with Down Syndrome

A Developmental Perspective

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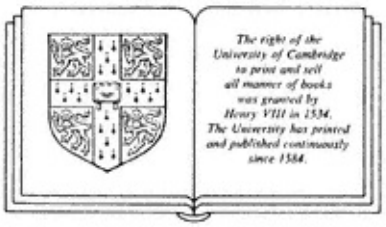

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Preface

Beginning with the writings of Esquirol (1838), who provided the first description of a child with Down syndrome, and John Langdon Down, whose article "Observations on an Ethnic Classification of Idiots" (1866) describes some of the characteristics of the children who are his namesakes, theorizing and research about Down syndrome have proliferated in the scientific literature (see Booth, 1985; Gibson, 1978; and Rynders & Pueschel, 1982, for historical reviews). Although earlier explorations into unraveling the complexities of this condition were conducted primarily by researchers interested in its biomedical aspects, increasingly the study of Down syndrome has become a multidisciplinary endeavor. As the most common organic form of mental retardation, it is not surprising that Down syndrome has captured the attention of so many talented research investigators, educators, and clinicians.

Down syndrome occurs in approximately 1 out of every 600 live births (Hook, 1982), resulting in nearly 7,000 infants in the United States and 1,000 in the United Kingdom born with Down syndrome each year (Wishart, 1988). Down syndrome arises from an abnormality on chromosome 21 (Thuline & Pueschel, 1982). Chromosome pair 21 is the smallest of the 23 human chromosome pairs, possessing only about 1.5% of the total genetic material. Moreover, gene-mapping studies have revealed that only 10% to 20% of chromosome 21, the 21q 22 band on the long arm, is involved in Down syndrome (Patterson, 1987a, 1987b; Patterson et al., 1985; Smith, 1985). Nonetheless, the presence of an additional copy of this small chromosome exerts a profound impact on the developmental biology and psychology of Down syndrome (Benda, 1960, 1969; Breg, 1977; Cicchetti & Ganiban, in press; Cicchetti & Pogge-Hesse, 1982; Coyle, Oster-Granite, & Gearhart, 1986; Epstein, 1986; Lane & Stratford, 1985; Nadel, 1988; Penrose & Smith, 1966; Pueschel & Rynders, 1982; Smith & Berg, 1976).

Most cases of Down syndrome are caused by a meiotic non-disjunction of autosomal chromosome pair 21, resulting in the triplication of the entire 21st chromosome (Lejeune, Grautier, & Turpin, 1959). Approximately 95% of all cases of Down syndrome are characterized by this autosomal trisomy of the 21st chromo-

some; hence, in the literature Down syndrome has been referred to as "trisomy 21." In addition to the trisomy 21 type, there are two other cytogenetic subtypes of Down syndrome. Of these cases, 4% to 6% manifest translocation of a portion of chromosome 21 to another chromosome. Furthermore, 1% to 4% are a mosaic of normal and trisomic cells as an outcome of a non-disjunction that occurs during early embryogenesis (Thuline & Pueschel, 1982).

Major congenital anomalies, involving virtually all senses and each organ system, are present in one-half of infants born with Down syndrome (see Cicchetti & Beeghly, Chapter 2, this volume; Coyle et al., 1986; Lane & Stratford, 1985; Pueschel & Rynders, 1982; Smith & Berg, 1976). Of all individuals with Down syndrome, 1% develop leukemia, a rate between 20 and 50 times greater than that found in the normal population (Coyle et al., 1986). While 30 to 40 years ago only 50% of all infants with Down syndrome survived infancy, recent medical advances (particularly in cardiology and immunology) have resulted in a significant increase in the life expectancy of persons with this condition. Because many individuals with Down syndrome now survive into their 50s and 60s, the prevalence of Down syndrome has increased by several hundredfold during the course of the 1980s (Coyle et al., 1986; Hook, 1982; Wishart, 1988). In concert with these biomedical breakthroughs, the findings of recent psychological and educational investigations have led to increased expectations for persons with Down syndrome (Lane & Stratford, 1985; Pueschel, Tingey, Rynders, Crocker, & Crutcher, 1987; Rynders, Spiker, & Horrobin, 1978).

Important discoveries in neurobiology likewise have occurred within the past several decades (Coyle et al., 1986). Among the most notable is the finding that the majority of adults with Down syndrome develop pervasive Alzheimer-like brain pathology by their mid-30s. Importantly, only about one-third of all individuals with Down syndrome manifest symptoms of a clinically diagnosed dementia (Thase, 1988). Thus, future investigations of the mechanisms that underlie the uncoupling of the Alzheimer-like neuropathology and dementia in Down syndrome could contribute greatly to the understanding of the ontogenesis and pathogenesis of Alzheimer's syndrome.

As a direct consequence of these advances and changes, it has become increasingly critical to bring a life-span developmental perspective to bear in

planning medical, educational, and psychological interventions for children with Down syndrome and their families. During the course of the past several decades, investigators have begun to employ a developmental perspective in their studies of infants and children with Down syndrome. Especially throughout the 1970s and 1980s, major advances have been achieved in several areas of the developmental psychology of Down syndrome, most prominently in the perceptual, social, emotional, cognitive, and representational domains. Contemporaneously, developmental neuroscientists have made significant progress in unraveling many of the genetic, biochemical, and neurophysiological mysteries of Down syndrome. As a consequence, we are approaching the point where a comprehensive developmental theory of Down syndrome may be formulated.

A unifying theme of this volume involves the premise that an organismicorganizational developmental approach can be applied to the study of any discipline (e.g., education, neuroscience, pediatrics, psychiatry, and psychology) and to any population, normal or otherwise atypical. Specifically, the contributors to this book believe that despite the constitutional and biomedical problems described earlier, children with Down syndrome can be studied through this developmental framework. The particular thesis espoused is that the study of children with Down syndrome from such a developmental perspective will increase our knowledge of this condition, thereby minimizing the generation of stereotypes about it and maximizing our understanding of Down syndrome from a lawfully organized viewpoint. Conversely, through the study of Down syndrome from a developmental perspective, advances can occur in the formulation of a truly integrative theory of human development. Such a study also highlights the respective roles of biological, perceptualcognitive, linguistic, social, emotional, and representational factors in the developmental process. Furthermore, the study of Down syndrome permits us to identify alternative pathways or processes to adaptive and maladaptive outcomes. Finally, to the extent that we now possess a great deal of information on the genetic, biochemical, physiological, and psychological aspects of infants and children with Down syndrome, the developmentalist interested in this condition is in the unique and enviable position of examining the interaction between heredity and environment on the developing organism.

Advances such as these also can be instrumental in informing and guiding methods of intervention. Efforts influenced by developmental principles in psychology, education, and biology are more likely to result in informed knowledge about how best to help children with Down syndrome and their families. Too often parents of these children are presented with inaccurate information that builds false hopes and ultimately causes grave disappointment. Adherence to the developmental perspective should lead to the implementation of timed, guided, and appropriate forms of intervention.

In this volume, authors have contributed theoretically informed chapters on the state of current knowledge of the various domains of psychological development in infants and children with Down syndrome. Where the data base permits, the biological systems that play important roles in the ontogenesis of these psychological domains are discussed. Additionally, a comprehensive treatment of temperament in Down syndrome and its relation

to biological and psychological processes, is provided. Furthermore, interventions for children with Down syndrome are reviewed from a developmental perspective. Finally, throughout the volume, suggestions for future research are suggested. We believe that many exciting ideas have been generated by the contributors to this volume and eagerly anticipate the new wave of multidisciplinary developmentally guided research on Down syndrome across the life-span.

We would like to explain our decision to employ "Down" rather than "Down's" throughout this volume. For several decades, parents of children having this condition have advocated that the term *Down syndrome* be utilized because of their

concern that their children were being viewed as simple extensions of the syndrome they possessed. By adopting the terminology of the parents, we are expressing our support for the individuality of the children, despite their shared syndrome. We believe that agreeing with the philosophy of these parents, as well as that of the many scientists in the area who also have begun to use this term, is essential.

On a more personal note, we would like to thank all of the children with Down syndrome and their parents who have participated in our research in Pittsburgh, MinneapolisSt. Paul, and Boston over the years. We feel privileged to have gotten to know them (some extremely well), and feel that our lives are richer as a result. We wish them continuing happiness in their lives together. To them, as well as to all children with Down syndrome and their families, we dedicate this book.

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