Selected References
(Note: A selected list of current papers on Down syndrome is published in each issue of Down Syndrome Quarterly. This feature is edited by David Smith MD. (DS Center of Wisconsin in Milwaukee WI.)

- References marked with an asterisk are those added to the 1999 revision of the Health Care Guidelines

A. Overview

Giesinger, C., for the Canadian Down Syndrome Society. Annotated bibliography of journal articles on Down syndrome for parents and primary caregivers. Calgary, AB: CDSS, no date. (Telephone 403-270-8500)


B. Adult Health


C. Other Checklists and Protocols

Pediatrics, 93, 855-858.


D. Specifically for Females


E. Anesthesia


F. Audiology


G. Cardiology


II. Communication


I. Dental


J. Development


K. Ear, Nose and Throat/Sleep Apnea


L. Education


M. Endocrinology


N. Nutrition & Feeding/Gastrointestinal


O. Genetics and Prenatal Screening


P. Growth


See also, Section Z. "Internet Resources," below, for URL for Growth Charts

Q. Gynecology


R. Hematology/Oncology


S. Immunology


T. Longevity, Mortality and Long-term outcome


U. Neurology (See X. Psychiatry, Neurology, and Developmental Biology, below.)

V. Ophthalmology


W. Orthopedics & Atlanto-Axial Instability


*Y. Alternative Therapies


**Z. Internet Resources

1. National Organizations

   National Down Syndrome Society: www.ndss.org

   National Association for Down Syndrome: www.nds.org

   National Down Syndrome Congress: www.members.carol.net/~ndscl

2. Journals

   Down Syndrome Quarterly, www.denison.edu/dsq


   Disability Solutions. www.disabilitysolutions.org

3. General Information

   1999 Health Care Guidelines: www.denison.edu/dsq


   This is a comprehensive list of health information about DS, including lists of DS Clinics in the US and abroad. You will find a listing of Current Ongoing Research in Down syndrome from the US Public Health Service database. This award-winning site, authored by Dr Len Leslin, has articles on common health concerns, such as constipation and gastroesophageal reflux, as well as monthly abstracts from the scientific literature.
Book Review

Barry M. Mitnick, Ph.D.
Book Review Editor

Adolescence and Down Syndrome


REVIEWED BY: Barry M. Mitnick, Ph.D.
University of Pittsburgh

Here is a tale of two high schools in the same county: Three delightful young men with Down syndrome, 18-19 years old, attend them. At FC High School, considered one of the best in the U.S. for college-bound students, Ed went through the graduation ceremony with his peers; he will get his actual diploma when he leaves the school system at age 21. His non-DS friends came to his graduation party; so did an assistant principal and his guidance counselor.

At the same school, Jeff is a junior; school administrators expect that Jeff, too, will attend his graduation ceremony next year. His parents will retain school district support for Jeff until age 21, and that is no problem here. Ed was the pioneer in this school district; the district adopted a benign policy of supported inclusion as a result of his mother’s efforts a few years earlier.

The academic experience of a student with Down syndrome is still mixed at this high school, despite clear policy at the district level: There are some superb teachers, both special and regular education, who go above and beyond in working with kids like Ed and Jeff; there are also some special education teachers who say they have no idea how to teach kids like Ed or Jeff or who simply neglect to do the necessary work on things like math programs and transition plans. Opportunities for social inclusion in the school’s activities could be better, though Ed is a popular manager for the football team.

In contrast, at WH High School, Scott was given the choice of attending his graduation ceremony, receiving his actual diploma and losing all future school system support, or deferring his attendance at graduation (and actual graduation, of course) to age 21 and retaining that support. The media covered his case, with editorials in the major newspaper supporting Scott’s desire to attend graduation with his peers. The school board was unmoved, and Scott had his “graduation” at home, with friends and family attending. The case has attracted national attention, and continues in litigation. Scott’s parents say that the fact that it occurred at all is remarkable; in their view the district accommodated or attempted to accommodate all their requests during Scott’s schooling, until the issue of attending graduation arose.

The experience of the adolescent with Down syndrome in the United States, even in school settings as relatively benevolent as those at FC and WH High Schools, remains mixed. Even where the basic legal battles have been won, more subtle and sometimes equally critical issues remain. Though provided with services, Scott was clearly considered a student without peers, not really a member of the class that contained his friends. Not separate, perhaps, in his school career, but clearly unequal, at least in the opportunity to celebrate the basic rite of school passage.

Unlike Jeff and Ed, students without Down syndrome at FC High School will never be told by any of their teachers that they have no idea how they should be taught. They will never experience teachers whose neglect of their duties leads to outside consultants being brought in to write essential parts of their education plans. And they will be lucky to enjoy teachers who know them as well and work as hard in their interests as some teachers have for Jeff and Ed. That is another kind of inequality, of specialness; one whose positive benefits should extend to all, not just our “special” children.

Piecemeal Knowledge and Complex Systems

The problems of providing optimal care and educational opportunities for adolescents like Ed, Jeff, and Scott are complex, and cannot be solved piecemeal. As Pueschel and Šustrová argue in their comprehensive reference work on Adolescents with Down Syndrome, the advance of knowledge has made possible “a better understanding of the total person with Down syndrome” (p. xix). As a “total,” the person with Down syndrome must be understood as more than a set of issues or concerns, and more than a compilation of academic subspecialties. To specific knowledge must be added general management; the system itself is far more complex than any one of its also complex parts. We can sketch the figure with dots, but we must also join those dots with smooth line.

Indeed, when approaching a terrific source book like this one, jam-packed with useful summaries of the literature and observations about enhancing the experience of adolescence in children with Down syndrome, one can tote up the impressive contributions that have been made to our knowledge about DS. This represents the “normal science” in the systematically-derived generalizations about the phenomena of adolescence in Down syndrome. But after we tote it up, accumulate it, bind the pieces together in this volume, do we have something greater than that? We have a rich collection of
specialized knowledge, but do we have a sure understanding of how to enhance adolescence in individuals with Down syndrome?

In a way, this is not a fair question. We have been so starved for knowledge about Down syndrome. As the field has progressed, it has been as if it has aged with its subjects: It was not long ago that virtually all of the collections like this emphasized very young children. Buckley and Sacks (1987) stood out not only for its quality but for its isolation in the literature. No doubt in a few years we will see a profusion of similar collections concerning adults with Down syndrome; it would be most welcome.

But now that we recognize the flourishing of sub-areas of knowledge about Down syndrome, we must ask if and how it makes a difference for Ed, Jeff, and Scott. This is a body of knowledge about one syndrome, and it applies to whole bodies of real kids. How in the world do you pull the pieces together and make them work in real-life settings? Does a book like this belong up there on the shelf in the Essential Library about Down syndrome? Yes, it does. But if so, how and when do you know to take it down and use it?

Thanks to Mary Coleman's original model and the creative persistence of savants like Sieg Pueschel, Bonnie Patterson, Len Leshin, Bill Cohen, George Capone, and, thankfully, a number of others equally distinguished, we have created a network of centers. Most often at the core of such a center is an individual or two whose job involves, in part, mastering big chunks of the burgeoning knowledge base. Most are physicians, and emphasize the medical knowledge areas. Even so, one of their chief tasks is to recognize when and to whom referrals are necessary. In essence, the knowledge collected serves to recognize the potential for problems that are dealt with elsewhere. Has the adolescent been evaluated for a potential atlantoaxial problem? Is the weight gain due to hypothyroidism? But it is in orthopedics or endocrinology that the potential problem is evaluated and actually dealt with.

So who really takes care of these kids? Well, of course, their parents. But none of us can truly master what is in a book like this. Ultimately, we do referrals, too: Referrals to transition coordinators, speech therapists, lifeskills teachers, and Down syndrome clinic directors. Perhaps the key pattern distinguishing the lives of kids with and without Down syndrome is the degree to which their lives are influenced by agents. Like all agency relationships, those in service to adolescents with Down syndrome have their faults; in the real world we rarely get perfect agents. Hence we face a double dilemma: dependence on a host of imperfect agents tied together in a system that nobody masters. So once we compile the master pile of knowledge one of our chief chores will be ensuring that its agents/carriers do with it what we want; knowledge in the absence of effective implementation is not very valuable. And even if our agents are perfect, we must devise means to ensure that these best efforts do not work at cross-purposes, that they create, collectively, the finest quality of life for Ed, Jeff, and Scott.

Adolescence and Down Syndrome

Pueschel and Šustrová have done an admirable job of collecting (and, in some cases, writing) twenty-five solid, valuable essays about many of the key issues concerning adolescence and Down syndrome. The book is well-edited. For example, brief cases are presented uniformly in boxes, and each chapter has a set of clearly-written conclusions that summarize its content. There are many excellent tables. The book also has a useful index and an appendix that features a helpful list of agency resources, complete with phone and fax numbers.

The first section of the book consists of six well-done essays on health and physical development. Perhaps, some day, the quality of life itself, rather than the steps necessary to support it, will have first billing. There is a logic to reading about the medical precursors first—most collections begin this way. We may find good health essential, and work first to achieve it, but we must remember that ultimately what we seek are happy, fulfilling lives.

Pueschel and Šustrová lead off with a piece on general health issues and medical care, and follow with one on nutritional concerns. They note that “the vast majority of adolescents with [Down syndrome] usually enjoy good health” (p. 9), a fact that is easy to overlook among the long discussions about conditions that appear to be a bit more prevalent in DS that typically dominate works like this. Pueschel and Šustrová note the existence of health care checklists (they do not cite the one that appears in Down Syndrome Quarterly, perhaps because this chapter may have been prepared close to the time that DSQ commenced). This section is more of a medical overview than a detailed discussion; it focuses on what areas the physician encountering an adolescent with DS should investigate and, later, track.

Both the apparent tendency of young people with Down syndrome to become overweight, and the existence of claimed therapies based on various nutrients, make nutrition a good subject for a second chapter. Šustrová and Pueschel offer a good, practical analysis of the problems of obesity, note an uncertain potential for celiac disease, and summarize some of the vitamin/mineral therapies. They do not address the recent controversies over piracetam.

Christine Cronk’s chapter on growth, which focuses usefully on measurement, including the evaluation of height (and of height vs. weight), logically follows. She does not address the controversies over growth hormone and DS.

Pueschel then provides a brief chapter on adolescent development and sexual maturation, which includes basic descriptive material on development that is not often found in the article collections like this. The fifth chapter, by Lawson and Elkins, addresses gynecologic concerns, noting both the basic similarity to non-DS females and a variety of issues special to DS.

The last chapter in this section, again by Pueschel and Šustrová, looks at some medical conditions selected to be especially relevant for adolescence. These include dermatologic disorders, ophthalmologic conditions, ENT concerns, cardiac issues, thyroid dysfunction, and atlantoaxial instability. This chapter is quite well-done, highlighting many important and/or frequently-encountered conditions.
To take a small example, how many pediatricians/internists will recognize the frequency of follicular infections as characteristic and pay attention to their treatment?

The second section of the book deals with behavioral, psychologic, and psychiatric issues. It leads off with a chapter by Jeanie P. Edwards on growing into a social-sexual being. She emphasizes the importance of social skills training and competence building in training about relationships and sexuality. Meshing well with this is a chapter by Powers and Sikora on promoting adolescent self-competence. They identify important approaches to promoting self-competence, though most of the discussion could apply to any adolescent, not just those with DS.

Robert Hodapp’s chapter is a nice analysis of the frequent disconnection between cognitive level and practical functioning: measures of cognitive achievement are often not particularly informative in regard to display of everyday skills. Robin Chapman looks at language development, noting some particular issues in DS, and challenges the conclusions of some earlier work that suggested that there may be a limit or ceiling in the ability to produce ever longer, more complex sentence patterns as the individual ages.

Cuskelly and Gunn present an analysis of behavior concerns in DS, concluding that most adolescents with DS do not present behavior management problems. They discuss the management of those issues that do arise from time to time (e.g., nonproductive persistence or perseveration). Beverly Myers offers a useful chapter on psychiatric disorders, noting the types and prevalence of such disorders. Depression occurs more in young people with DS than in people with mental retardation of different origin. She notes that, in general, however, most psychiatric disorders are not more common in DS than in comparison groups.

The third section of the book focuses on education.

Falvey, Rosenberg, and Falvey provide an excellent descriptive overview of current practices and strategies or programs said to promote successful inclusion. It is not specific to DS. A problem with this chapter, however, is its lack of attention to implementation issues. These programs do not always work well in practice, and the reader is given no sense of their limitations, or the manipulations that real-world settings have given to them. Block scheduling, for example, has both advantages and disadvantages for the special student, and we see nothing of the down side. Attention to problems in adaptation of materials, teacher competence, teacher implementation, proper use of cooperative learning modes, and so on, is missing. Even where the balance of experience might suggest a worthy program (which is likely the case for block scheduling, for example) we hear nothing of that practical balance.

Bud Fredericks provides a thoughtful discussion of alternative patterns of secondary and post-secondary curriculum. This should be especially valuable to those parents or professionals whose secondary school settings perceive only limited alternatives in the opportunities available to special students.

Joan Tanenhaus provides a useful description of basic issues in the use of the computer. The article is an extended checklist of things to think about rather than a particular learning approach implemented via the computer. It includes a long list of suggested software.

Getzel, Flippo, Wittig, and Russell present a chapter on postsecondary education, noting the need to identify appropriate participants in planning as well as the process of identifying appropriate postsecondary programs.

Salome Heyward provides a good review of legal mandates and protections. The very presence of a chapter like this suggests that the real world of adolescent education and service provision is not nearly as seamless as some of the other chapters in this book seem to suggest.

The fourth section of the book addresses life in the workplace.

Luecking and Fabian summarize some best practices in transition planning. They offer seven sensible strategies that may promote better, more successful job placements. Wehman, Parent, Unger, and Gibson examine supported employment, sketching its major features, advantages, some key aspects that promote success, and some suggestions on gaining access to such services. This valuable chapter has several cases and a table of sample positions (and support needs) that illustrate its points.

The chapter by Helm, Kiernan, and Miranda on opportunities for employment examines what has in recent years finally become "a realistic option" (p.267). They review the nature and advantages of person-centered planning and of some actions that can promote more successful job placement and adaptation. They also recognize and discuss some important concerns about sustaining employment and the individual’s experience with it.

The fifth and last section of the book addresses life in the community.

Debra Lanseth takes a look at the role of the family, or perhaps more properly, at the multiple and essential roles that family plays as advocate, service coordinator, evaluator, direct service provider (e.g., in teaching basic life skills), communication and self-esteem developer, and role model. She also considers, among other areas, risks that parents must evaluate and, ultimately, take in promoting their children’s independence and integration in the community.

In an interesting but too-brief chapter, Nisbet, Crowley, and Crowley note the importance of full inclusion in the community and provide a list of steps that could lead to this. This is a critical issue: We have focused so intensely on the early years that we tend to forget that most of life comes later. Inclusion in the community deserves a longer, more systematic exploration.

One component of community inclusion is involvement in leisure-time activities. Heyne, Schleien, and Rynsers carefully and systematically examine the role of recreation, describing strategies to promote participation, including assessment of needs and interests, selection of activities, task analysis of the activity and the design of adaptations, the use of behavioral teaching methods, and the evaluation of the participation. Several cases are presented. Songster, Smith, Evans, Munson, and Behen review one of
the most widespread and successful recreational programs for special needs individuals, Special Olympics.

The book concludes with a chapter by Julie Racino on youth and community life. Its content is in general not specific to Down syndrome. The focus is on public policy frameworks and perspectives, as well as particular programs or services, and types of programs, that promote and support success in living in the community. She lists seven frameworks that have guided program development (for example, a “capability-autonomy” perspective that rests on notions of social justice and personal autonomy rather than on a “minority group” framework that focuses on entitlement to special accommodations (pp. 362-363)). She looks at the objectives of providing personal assistance to achieve better opportunities and the ability to realize personal goals, at public policy approaches toward housing, recreation, education, and employment, and at the roles of personal assistance and of systems of support. She also lists some person-centered mechanisms to produce change in such areas as services and community support. This chapter reads more like an inventory drawn from longer works than a useful, critical analysis and evaluation of alternative frameworks.

Of course, as a whole this book is far more than an inventory of observations and generalizations. It stands as the major existing source book on adolescence and Down syndrome. It lays out the chief areas of our knowledge.

In the future, besides deepening and expanding our knowledge, however, we must also look to the practical question of how to make our systems of care and inclusion work better. If there is to be no systematic, central master of this formidable mass of learning, we need to craft systems that serve masterfully. How can we know all this, yet fail to provide Ed, Jeff, and Scott with all that they deserve in school and the community? How can we manage pervasive networks of agents? It would be a grand and sad irony, if the struggle to construct vessels of understanding in this long-neglected realm is successful, only to find their essential passengers adrift at sea.

References


News from the Down Syndrome Medical Interest Group (DSMIG)

William I. Cohen, M.D.
Bonnie Patterson, M.D.
Co-Chairs

Mission The Down Syndrome Medical Interest Group (DSMIG) was founded in early 1994 with the express purpose of serving as a forum for professionals addressing aspects of medical care of persons with Down syndrome. DSMIG wishes to promote the highest quality care for children and adults with DS; 1) by fostering and providing professional and community education; 2) by disseminating tools for clinical care and professional support; such as the Health Guidelines for Individuals with Down Syndrome; 3) and by engaging in collaborative clinical research regarding issues related to the care of individuals with Down syndrome.

For further information, contact either co-chair: Bonnie Patterson at 513-559-4691 or Bill Cohen at 412-692-6546.

If you are interested in being added to our mailing list, please send your name, professional title, agency, address, telephone number, fax number, and email address (if any) to William I Cohen MD, Down Syndrome Center, Children’s Hospital of Pittsburgh, 3705 Fifth Avenue, Pittsburgh, PA 15213. (412-692-6546; fax 412-692-5679; email: cohenh@chplink.chp.edu).

News From DSMIG

Thursday, November 11, 1999 is the date for the next DSMIG meeting, to be held at Children’s Hospital of Pittsburgh, PA. The meeting will be followed by a CME conference entitled “Down Syndrome: Clinical Guidelines, Research, and Education,” which will be held on Friday and Saturday, November 12 & 13, 1999. This conference marks the 10th anniversary of the Down Syndrome Center of Western PA. Scheduled participants include the following members of DSMIG: Drs David Patterson, Carl Cooley, George Capone, Ditza Zachor, Brian Chicoine, Dennis McGuire, among others. For further information about either program, contact Sue Haunty at Medical Relations, Children’s Hospital, 412-692-67507. (email: haunty@chp.com).

Tentative Agenda for DSMIG Meeting:
8:30 AM Breakfast
9:00 AM Welcome; Introductions
9:30 AM Health Care Guidelines 1999 Revision
New Initiatives-Development Guidelines for:

- Speech/Language
- Education
- Physical & Occupational Therapy
- Behavior Problems

10:30 AM Break
10:45 AM New Initiatives (continued)
Clinical Database
DSMIG Website

12:00 PM Lunch
1:00 PM Reconvene for Afternoon:
Presentations by Members
- Research Projects
- Short Case Reports
3:00 PM Break
3:15 PM Discussion of Clinical Cases
4:45 PM Summary: Discussion regarding future activities, including next meeting of DSMIG

Future Scientific Meetings and Conferences:


COMMUNICATION

Abstract. Examined the expressive vocabulary development in 130 children (aged 1-5 yrs) with Down syndrome. Families completed a short vocabulary development history and the MacArthur CDI/Words and Sentences test forms (L. Fenson et al, 1993). Findings are discussed in the areas of vocabulary growth, gender differences, signed and spoken vocabulary, referential and grammatical vocabulary, multilword combinations, and morphosyntactic structures. Results show that although there was continuous growth in expressive referential vocabulary from birth through 5 yrs, age 5 was found to be an important developmental marker for multilword combinations and grammatical vocabulary. Treatment applications for early intervention and extended to the birth-to-5yr period are discussed in detail. Appendices of the Vocabulary Development Questionnaire and resources to address expressive vocabulary use are provided. (c) 1999 APA/PsychNFO, all rights reserved. Reprinted with permission by the American Psychological Association. For further information contact psychinfo@apa.org.

DENTAL

Comment: Although the frequency of periodontal disease increased, the severity and progression was less than that previously reported.


EDUCATION & THERAPY


ENDOCRINOLOGY


COMMENT: A letter.

EPIDEMIOLOGY


COMMENT: More than 50% of infants with DS had associated defects that caused morbidity and mortality. N=87. Maternal smoking and consumption of more than three cups of coffee was significantly associated with an additional risk.

GASTROENTEROLOGY


COMMENT: A letter.


COMMENT: A combination of AGA and EMA tests resulted in 100% sensitivity and 100% negative predictive value.


GENETICS


GROWTH & DEVELOPMENT


HEMATOLOGY

COMMENT: A letter.


Comment: A letter.


CONTENTS

Health Care Guidelines for Individuals with Down Syndrome: 1999 Revision
(Down Syndrome Preventive Medical Check List)
Edited by William I. Cohen for the Down Syndrome Medical Interest Group ................. 1

Book Review (Adolescents and Down Syndrome:
A Review of Adolescents with Down Syndrome:
Toward a More Fulfilling Life by Siegfried M.
Pueschel and Maria Sustrova, Eds.)
Barry M. Mitnick ........................................... 16

News from the Down Syndrome Medical Interest Group
(DSMIG) William I. Cohen and Bonnie Patterson ......................... 19

References/Abstracts
David Smith ................................................. 20