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Evidence Report/Technology Assessment
Number 73

Criteria for Determining Disability in Infants and Children: Short Stature

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Prepared by:
Tufts-New England Medical Center EPC, Boston, MA

Investigators
Patricia Wheeler, MD
Karen Bresnahan, MD
Barbara Shephard, MD

EPC Staff
Joseph Lau, MD, Director
Ethan Balk, MD, MPH, Project Leader
Deirdre DeVine, M Litt, Project Manager
Mei Chung, MPH, Research Associate
Kimberly Miller, BA, Research Assistant

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Preface

The Agency for Healthcare Research and Quality (AHRQ), through its Evidence-Based Practice Centers (EPCs), sponsors the development of evidence reports and technology assessments to assist public- and private-sector organizations in their efforts to improve the quality of health care in the United States. The reports and assessments provide organizations with comprehensive, science-based information on common, costly medical conditions and new health care technologies. The EPCs systematically review the relevant scientific literature on topics assigned to them by AHRQ and conduct additional analyses when appropriate prior to developing their reports and assessments.

To bring the broadest range of experts into the development of evidence reports and health technology assessments, AHRQ encourages the EPCs to form partnerships and enter into collaborations with other medical and research organizations. The EPCs work with these partner organizations to ensure that the evidence reports and technology assessments they produce will become building blocks for health care quality improvement projects throughout the Nation. The reports undergo peer review prior to their release.

AHRQ expects that the EPC evidence reports and technology assessments will inform individual health plans, providers, and purchasers as well as the health care system as a whole by providing important information to help improve health care quality.

We welcome written comments on this evidence report. They may be sent to: Director, Center for Practice and Technology Assessment, Agency for Healthcare Research and Quality, 6010 Executive Blvd., Suite 300, Rockville, MD 20852.

Carolyn M. Clancy, M.D.
Director
Agency for Healthcare Research and Quality

Jean Slutsky, P.A., M.S.P.H.
Acting Director, Center for Practice and Technology Assessment
Agency for Healthcare Research and Quality

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Structured Abstract

Objectives. The evidence report provides a systematic review of the scientific evidence to answer three questions of whether short stature in a child 1) due to a medically determinable cause or 2) due to skeletal dysplasia may be associated with disability, and 3) whether decreasing growth velocity in a child with a chronic disease may serve as an indicator of severity of the disease.

Search Strategy. Systematic searches were performed for relevant articles in MEDLINE® from 1966 through February 2001, with updates through October 2001. Additional studies were identified from other databases, reference lists of review and primary articles, and from domain experts.

Selection Criteria. Eligibility criteria for study inclusion included: primary articles reporting original data on at least 10 children; in studies of children with short stature, data on association between height and functional ability or limitation; in studies of children with chronic diseases, association between a measure of severity of disease and either height or height velocity. Studies could be cross-sectional or longitudinal, prospective or retrospective, comparative or not.

Main Results. A total of 13,537 English language citations were reviewed. Of these, 825 articles were reviewed for inclusion. For Question 1, 31 studies met inclusion criteria; for Question 2, 31 studies; and for Question 3, 53 studies. Detailed data extraction was performed on these 115 studies.

Based on the reviewed articles, no severe functional limitations were found in children with short stature due to isolated short stature, growth hormone deficiency, multiple hormone deficiency, Turner syndrome, or Russell-Silver syndrome. The studies reviewed focused on intelligence, academic achievement, behavior, visual-motor perception, and psychomotor development. In each of these categories, children with short stature either had testing that was not significantly different from the controls or from the population mean, or if the testing were significantly poorer, it was generally within 1 standard deviation (SD) of the population mean.

Based on the articles reviewed, children with skeletal dysplasias were not at increased risk of having severe impairments in intelligence, academic achievement, or psychological outcome. There was an increased risk for delay in achievement of motor skills in children with achondroplasia and osteogenesis imperfecta, and decreased ambulation, range of motion and mobility in children with more severe forms of osteogenesis imperfecta. The results for hearing impairment, respiratory dysfunction and spinal curvature appear to indicate an increased risk for impairment in these three areas, but the studies were limited in the number of children evaluated and how the samples were selected.

The evidence from four conditions (congenital heart disease, juvenile rheumatoid arthritis, Crohn's disease and human immunodeficiency virus (HIV) infection) appear to indicate that a sustained decrease in linear growth velocity can be used as a marker of the severity of these underlying conditions. Evidence is less clear for asthma, atopic dermatitis, diabetes, ß-thalassemia, and chronic kidney disease. There was only one study each for sickle cell disease, congenital adrenal hyperplasia and cerebral palsy so it is difficult to draw conclusions for these conditions. No study addressed whether the process of having a decreasing height velocity was likely to be disabling.

Conclusions. No severe functional limitations were found in children with short stature due to growth hormone deficiency, multi-hormone deficiency, Turner syndrome, Russell-Silver syndrome or isolated short stature. These children generally scored within 1 SD of the population mean. Children with skeletal dysplasias were not at increased risk of having severe impairments in intelligence, academic achievement, or psychological outcome. Sustained decrease in linear growth velocity has been shown to be associated with the severity of congenital heart disease, juvenile rheumatoid arthritis, Crohn's disease, and HIV.

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