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Titolo	Advances in Gaucher disease : basic and clinical perspectives // editor, Gregory A. Grabowski
Pubbl/distr/stampa	London, England : , : Future Medicine Ltd, , 2013 ©2013
ISBN	1-78084-201-5
Descrizione fisica	1 online resource (266 p.)
Disciplina	616.3997
Soggetti	Gaucher's disease Gaucher's disease - Treatment Electronic books.
Lingua di pubblicazione	Inglese
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Livello bibliografico	Monografia
Note generali	Description based upon print version of record.
Nota di bibliografia	Includes bibliographical references at the end of each chapters and index.
Nota di contenuto	Title page; Copyright page; Contents; Foreword. Advances in Gaucher disease: basic and clinical perspectives; 1. Neuronopathic phenotypes of Gaucher disease; 2. Gaucher disease: a comprehensive review; 3. Gaucher disease: the neurological examination; 4. Relationship between Gaucher disease and parkinsonism; 5. Cytology of Gaucher disease; 6. Glycosphingolipid aspects of Gaucher disease lipidomics; 7. Molecular abnormalities in Gaucher disease tissues; 8. Crystal structure of the enzyme acid b-glucosidase; 9. Processing and maturation of human glucocerebrosidase 10. Properties of wild-type and mutant glucocerebrosidases 11. Diagnostic testing (enzyme and mutational analysis); 12. Imaging skeletal involvement in Gaucher disease; 13. Assessment of individual organs: other organs; 14. Biomarkers; 15. Current specific enzyme therapies; 16. Synthesis inhibition therapy for Gaucher disease; Index

2. Record Nr.	UNINA9910970651503321
Titolo	Advanced fibers for high-temperature ceramic composites : advanced materials for the twenty-first century // Committee on Advanced Fibers for High-Temperature Ceramic Composites, National Materials Advisory Board, Commission on Engineering and Technical Systems, National Research Council
Pubbl/distr/stampa	Washington, DC, : National Academy Press, 1998
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Edizione	[1st ed.]
Descrizione fisica	1 online resource (111 p.)
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Disciplina	620.1/4
Soggetti	Ceramic-matrix composites Ceramic fibers Heat resistant materials
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Nota di contenuto	CERAMIC FIBERS AND COATINGS -- Copyright -- Preface -- Acknowledgments -- Contents -- Acronyms -- Executive Summary -- APPROACH -- HIGH-TEMPERATURE CERAMIC FIBERS -- Non-Oxide Ceramic Fibers -- Oxide Ceramic Fibers -- FIBER COATINGS -- Coatings for Non-Oxide Fibers -- Coatings for Oxide Fibers -- RECOMMENDATIONS AND IMPACTS -- DISCUSSION OF PRIORITIES -- 1 Introduction -- APPROACH -- POTENTIAL CERAMIC MATRIX COMPOSITE APPLICATIONS -- COMPOSITE MATERIALS -- CERAMIC FIBERS AND COATINGS -- ENGINEERING REQUIREMENTS -- REPORT ORGANIZATION -- 2 Current and Future Needs -- IMPLEMENTATION OF NEW MATERIALS -- Glass Matrix Composites -- SiC Matrix Composites --

Oxide/Oxide Composites -- Inhibited Carbon Matrix Composites -- CERAMIC MATRIX COMPOSITE DESIGN AND LIFE PREDICTIONS -- CERAMIC MATRIX COMPOSITE APPLICATIONS AND REQUIREMENTS -- Energy Systems -- Aerospace Systems -- MANUFACTURING REQUIREMENTS -- Reliability and Reproducibility of Fiber Supplies -- Handleability and Processability -- IMPLICATIONS FOR FIBER PROPERTIES -- General Considerations -- Fiber and Interface Requirements -- 3 State of the Art in Ceramic Fiber Performance -- CANDIDATE FIBERS -- Comparison of Fiber Categories -- TEMPERATURE AND TIME DEPENDENCE OF PROPERTIES OF NON-OXIDE FIBERS -- Strength and Stiffness as a Function of Test Temperature -- Room-Temperature Strength and Modulus as a Function of Aging -- Thermal Conductivity (Diffusivity) and Electrical Conductivity (Resistivity) as a Function of Temperature and Aging History -- Creep Behavior of Non-Oxide Fibers and Effects of Heat Treatment, Oxidation, and Aging -- Rupture Behavior of Non-Oxide Fibers in Oxidizing and Non-Oxidizing Environments and the Effects of Heat Treatment and... -- Microstructural Stability -- TEMPERATURE AND TIME DEPENDENCE OF PROPERTIES OF OXIDE FIBERS. Strength and Stiffness as a Function of Test Temperature -- Room-Temperature Strength and Modulus as a Function of Aging History -- Thermal Conductivity (Diffusivity) and Electrical Conductivity (Resistivity) as a Function of Test Temperature and Aging... -- Creep Behavior of Oxide Fibers -- Microstructural Stability -- PERFORMANCE CHARACTERISTICS COMPARED TO PERFORMANCE GOALS -- RECOMMENDATIONS AND FUTURE DIRECTIONS -- 4 Ceramic Fiber Processing -- NON-OXIDE FIBER PROCESSING -- Preceramic Polymer Processing -- Microstructural Development -- OXIDE FIBER PROCESSING -- Chemical Processing -- Chemistry of Oxide Fiber Precursors -- Basic Aluminum Salts -- Polymeric Aluminosilicates -- Dry Spinning -- Pyrolysis -- Heat Treatment and Fiber Microstructure -- RECOMMENDATIONS AND FUTURE DIRECTIONS -- Non-Oxide Fibers -- Oxide Fibers -- 5 Materials and Microstructures -- OPPORTUNITIES FOR FIBER DEVELOPMENT -- POLYCRYSTALLINE OXIDES -- POLYCRYSTALLINE SILICON CARBIDE -- AMORPHOUS FIBERS -- RECOMMENDATIONS AND FUTURE DIRECTIONS -- 6 Interfacial Coatings -- COATINGS FOR NON-OXIDE COMPOSITES -- Carbon Coatings -- Composite Behavior -- Oxidation Behavior -- Boron Nitride Coatings -- Composite Behavior -- Oxidation Behavior -- Alternative Fiber Coatings -- Coating Compositions -- Oxidation Behavior -- Systems Approach to Inhibiting Oxidative Embrittlement -- Coating Processes and Vendors -- Chemical Vapor Deposition -- Vendors -- Alternative Approaches -- OXIDE FIBER COATINGS -- Porous Coatings and Porous Matrix Approaches -- Porous Coatings -- No Coating/Porous Matrix Approach -- Fugitive and Segregant Weakened Interfaces -- Fugitive Interfaces -- Segregant Weakened Interfaces -- Dense Oxide Fiber Coatings -- Nonreactive Oxides -- Layered Oxides -- Weakly Bonded, Nonlayered Oxides -- Coating Processes and Vendors -- Immiscible Liquid Coating Technique. Heterocoagulation Technique -- RECOMMENDATIONS AND FUTURE DIRECTIONS -- Non-Oxide Fiber Coatings -- Oxide Fiber Coatings -- Porous Coatings and Porous Matrix Approaches -- Fugitive and Segregant Weakened Interfaces -- Dense Oxide Fiber Coatings -- Coating Processes and Vendors -- 7 Cost Issues -- PRICE VS. COST -- TYPES OF COST -- Variable Costs -- Fixed Costs -- The Bridging Factor-Cycle Time -- GENERAL FIBER MANUFACTURING COSTS -- MANUFACTURING CERAMIC FIBERS -- OXIDES VS. NON-OXIDES -- FINDINGS -- Relationship between Price and Volume -- Is Fiber Price

Really the Key Issue? -- Improving Performance vs. Lowering Prices -- System Costs -- Implementation Costs -- CONCLUSIONS -- RECOMMENDATIONS AND FUTURE DIRECTIONS -- Improving Fiber Performance -- Reducing the Risk of Commercializing Ceramic Fibers -- Expanding the Potential Market -- 8 Recommendations and Future Directions -- ENGINEERING DATA -- FIBER COATINGS -- Coatings for Non-Oxide Fibers -- Coatings for Oxide Fibers -- DEVELOPMENT OF OXIDE FIBERS -- DEVELOPMENT OF NON-OXIDE FIBERS -- MANUFACTURING COSTS -- PRIORITIES -- References -- Biographical Sketches of Committee Members.

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Sommario/riassunto

High-temperature ceramic fibers are the key components of ceramic matrix composites (CMCs). Ceramic fiber properties (strength, temperature and creep resistance, for example)-along with the debonding characteristics of their coatings-determine the properties of CMCs. This report outlines the state of the art in high-temperature ceramic fibers and coatings, assesses fibers and coatings in terms of future needs, and recommends promising avenues of research. CMCs are also discussed in this report to provide a context for discussing high-temperature ceramic fibers and coatings.

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