Molecular mechanisms, clinical manifestations, and new treatments

Monogenic Hyperinsulinemic Hypoglycemia Disorders

Editors

Charles A. Stanley
Diva D. De León

In this volume of Frontiers in Diabetes dealing with the molecular basis of monogenic disease of beta-cell insulin regulation, renowned experts provide in-depth descriptions of the most recent advances in genetic defects that cause hyperinsulinemia. These disorders comprise the most important form of hypoglycemia in infants and children and are associated with a high risk of mortality, including seizures and severe brain injury. The discovery of eight different genetic loci involved in congenital hyperinsulinism has led to greatly improved methods of diagnosis and treatment. Several new treatments are highlighted, such as 18F-DOPA PET scans for preoperative localization of focal hyperinsulinism, as well as potential new treatments, such as green tea extract for genetic hyperinsulinism disorders. Researchers will discover information for clinical diagnosis, management, and new treatments for infants and children with congenital hyperinsulinism. Readers will discover how genetic hyperinsulinism disorders provide novel insights into the basic mechanisms regulating insulin secretion not only in diabetics, but also in healthy humans.

Contents

Preface: Stanley, C.A.; De León, D.D.
Historical Perspective on the Genetic Forms of Congenital Hyperinsulinism: Stanley, C.A.; Matschinsky, F.M.
Biochemistry and Physiology of the ATP-Sensitive Potassium Channel Hypoglycemia: De León, D.D.; Stanley, C.A.
Phenotypic Significance of Dihydropyridine Receptor Defects in Congenital Hyperinsulinism: De León, D.D.; Stanley, C.A.
Role of Incretin Hormones in Hyperinsulinemic Hypoglycemia: Stanley, C.A.; De León, D.D.
Amino Acid-Stimulated Insulin Secretion: The Role of the Glucagon-like Peptide-1 Receptor: Canfield, S.; Matschinsky, F.M.; Stanley, C.A.
Amine Oxidase Inhibitors: A Role in Congenital Hyperinsulinism: Remedi, M.S.; Nichols, C.G.
Amino Acid Stimulation: Symposium: Otonkoski, T.; Meissner, T.
Molecular Defects of ATP-Sensitive Potassium Channels in Congenital Hyperinsulinism: Shyng, S.L.; Bushkin, J.D.; Pretot, C.E.; Zhao, Q.
Localization of a Focal Lesion of Congenital Hyperinsulinism Imaging and Surgery: Jost, L.; Adler, N.J.
Role of Incretin Hormones in Hyperinsulinemic Hypoglycemia: De León, D.D.
Glutamate Dihydropyrimidinase: Structure, Regulation, and Its Role in Islet Hormone Secretion: Smith, T.Z.

Conventional Hyperinsulinism Due to Activating Mutations of Glucokinase: The Hyperinsulinism/Hyperammonemia Syndrome: Stanley, C.A.
Granule Membrane Protein 2: The Role of a Novel Component of the Secretory Pathway in Congenital Hyperinsulinism: Verkarre, V.; Rossignol, S.; Aigrain, Y.; de Lonlay, P.
Molecular Defects of ATP-Sensitive Potassium Channels: Matschinsky, F.M.; Stanley, C.A.
Amino Acid-Stimulated Insulin Secretion: The Role of the Glucagon-like Peptide-1 Receptor: Canfield, S.; Matschinsky, F.M.; Stanley, C.A.
Molecular Genetics and Pathophysiology of Congenital Hyperinsulinism Caused by Short Chain 3-Hydroxyacyl-CoA Dehydrogenase Deficiency: Mokere, A.; Herpigel, G.
Exercise-Induced Hyperinsulinism: A Failure of Monocarboxylate Transporter 1 Expression Silencing: Otonkoski, T.; Meissner, T.
Incretin Hormones Due to Mutations of Uncoupling Protein 2: The Role of Mitochondria in the Pathogenesis of Congenital Hyperinsulinism: Matschinsky, F.M.; Stanley, C.A.
Glutamate Dihydropyrimidinase: Structure, Regulation, and Its Role in Islet Hormone Secretion: Smith, T.Z.

Author Index
Subject Index

Endocrinology; Pediatrics; Pediatric Endocrinology, Diabetes, Neonatology

M. Porta, F.M. Matschinsky

Editors

Diva D. De León
Charles A. Stanley

Frontiers in Diabetes, Vol. 21
Series Editors: Frentz, M. (Tartu), Matschinsky, F.M. (Rotterdam)
ISSN 0251-5342 / e-ISSN 1662–2995

210 Cognitive and Neurological Outcome at the Age of 5–8 Years of Premature Infants with Post-Hemorrhagic Ventricular Dilatation Requiring Neurosurgical Intervention

211 Postnatal Rosiglitazone Administration to Neonatal Rat Pups Does Not Alter the Young Adult Metabolic Phenotype
Truong, N.C.; Abbasi, A.; Sakurai, R.; Lee, W.N.P.; Torday, J.S.; Rehais, V.K. (Torrance, Calif.)

212 Is Octreotide Treatment Useful in Patients with Congenital Chylothorax?
Harvey, M.; Moon, C.F.; Antonius, T.A.J. (Nijmegen)

213 Efficacy and Safety of a Tight Glucose Control Protocol in Critically Ill Term Neonates
Verbruggen, S.C.; Landzaat, L.J.; Reins, I.K.M. (Rotterdam), van Goudoever, J.B. (Rotterdam/Amsterdam), Joosten, K.F.M. (Rotterdam)

214 Novel Insights from Clinical Practice

Gorcani, A.; de Carolis, M.P.; Piastra, M.; Bersani, I.; Padov, M.; Stival, E.; Tempera, A.; Romagno, C.; Conti, G.; De Rosa, G. (Rome)

216 Systematic Review and Meta-Analysis

183 Surfactant Lavage Therapy for Meconium Aspiration Syndrome: A Systematic Review and Meta-Analysis

217 Sources of Neonatal Medicine

159 Thrush – Nightmare of the Foundling Hospitals
Olshaban, M. (Berlin)

166 Noninvasive Assessment of the Early Transitional Circulation in Healthy Term Infants
Popat, H.; Khakpour, M. (Sydney, N.S.W.)

172 Recognition, Diagnosis and Treatment of Meconium Obstruction in Extremely Low Birth Weight Infants

179 First Day of Life Reference Values for Plast Variability Index in Spontaneously Breathing Term Newborns
Latini, G. (Brindisi/Pisa), Dippala, L. (Pisa); De Felice, C. (Vitera)

192 Differential Hemodynamic Effects of Levsimendan in a Porcine Model of Neonatal Hypoxia-Reoxygenation
Esch, J.; Jynct, C.; Manouchihi, N.; Lee, T.F.; Li, Y.-Q.; Bigun, D. (Edmonton, Alta.); Vento, M. (Valencia); Cheung, P.Y. (Edmonton, Alta.)


206 Cognitive and Neurological Outcome at the Age of 5–8 Years of Premature Infants with Post-Hemorrhagic Ventricular Dilatation Requiring Neurosurgical Intervention
Adipose Tissue Development
From Animal Models to Clinical Conditions

Editors
Claire Levy-Marchal
Luc Pénicaud

Contents
Preface
Levy-Marchal, C.
Human Lipodystrophies: Genetic and Acquired Diseases of Adipose Tissue
Capron, J.; Mogli, J.; Caron-Delbrock, M.; Leguilloux, G.; Azéma, K.; Benit, V.; Lecointe, O.; Barcelo, J.-P.; Vigneau, C.
The Emergence of Adipocytes: Laharrague, P.; Castella, L.
Adipose Tissue and the Reproductive Axis: Biological Aspects: Noonan, G.J.; Barker, C.R.
Unraveling the Obesity and Breast Cancer Links: A Role for Cancer-Associated Adipocytokines? Diérot, B.; Rael, L; Kornaros, G.; Weitz; Muller, C.
Early Determinants of Obesity: Ong, K.K.
Metabolic Syndrome in Childhood — Causes and Effects: Weiss, K.
Pathophysiology of Insulin Resistance in Small for Gestational Age Subjects: A Role for Adipose Tissue? Bellrand, J.; Menes, T; Levy-Marchal, C.
The Neural Feedback Loop between the Brain and Adipose Tissue: Pénicaud, L.

With some of the greatest advances from developmental endocrinology to clinical research being observed as a direct result of close collaboration between basic and clinical investigators, ‘Hormone Research in Paediatrics’ facilitates the interchange of ideas by pooling findings at all levels and from all branches side by side in a single source. Original research papers provide a background of experimental data on the pathology, cyogen, histology, biochemistry, pharmacology and regulation of hormones. This information is complemented by authoritative reports drawing on clinical experiences concerning the many diagnostic and therapeutic procedures and possibilities. In particular, such topical problems as growth, diabetes, obesity, sexual development and tumours of endocrine origin are covered in depth. As a bonus for subscribers, the journal also features frequent supplement issues.

More information at www.karger.com/hrp
• Pay-per-view and Subscriber Access to Full Text
• Full Table of Contents
• Full Editorial Board
• Free Abstracts and Selected Articles
• Online Sample Issue
• Submission/Guidelines for Authors
• Subscription Details
• Free Alert Service
• Online Library Recommendation

Adipose Tissue Development
From Animal Models to Clinical Conditions

The Neural Feedback Loop between the Brain Pathophysiology of Insulin Resistance in Metabolic Syndrome in Childhood — Causes and Effects

Nowadays, adipose tissue is not only regarded as an organ of storage related to fuel metabolism but also as an endocrine organ involved in the regulation of insulin sensitivity, lipids and energy metabolism.

These proceedings cover the nervous regulation of both white and brown adipose tissue mass. Different physiological parameters such as metabolism (lipolysis and thermogenesis) and secretory activity (leptin and other adipokines) are reviewed. The plasticity of adipose tissue (proliferation, differentiation and apoptosis) showing the presence of a neural feedback loop between adipose tissue and the brain, which plays a major role in the regulation of energy homeostasis, is discussed.

Merging basic knowledge and various clinical conditions, this thorough review is of great interest to both scientists and physicians, in particular pediatricians, interested in obesity, endocrinology and nutrition.

Please send to:

Name/Address:

E-Mail orders@karger.ch

www.karger.com/hrp

More information at www.karger.com/hrp

A qualified source of experimental data and informative clinical reports

HORMONE RESEARCH IN PÆDIATRICS
From Developmental Endocrinology to Clinical Research

Editor
P. Czernichow, Paris

Associate Editors
S. Bermon, Parma
F. Chiarelli, Chieti
W.S. Coutfeld, Auckland
M.T. Dattani, London
O. Hart, Lübeck

With some of the greatest advances from developmental endocrinology to clinical research being observed as a direct result of close collaboration between basic and clinical investigators, ‘Hormone Research in Paediatrics’ facilitates the interchange of ideas by pooling findings at all levels and from all branches side by side in a single source. Original research papers provide a background of experimental data on the pathology, cyogen, histology, biochemistry, pharmacology and regulation of hormones. This information is complemented by authoritative reports drawing on clinical experiences concerning the many diagnostic and therapeutic procedures and possibilities. In particular, such topical problems as growth, diabetes, obesity, sexual development and tumours of endocrine origin are covered in depth. As a bonus for subscribers, the journal also features frequent supplement issues.

Selected contributions
Validation of Bone Age Methods by Their Ability to Predict Adult Height: Thoburn, R.M.; Moon, J.C.; Keshel, J.; Langley, M.B. (Chicago), Jouve, A.G. (Paris), Martin, M.B. (Düsseldorf)
Pediatric Graves’ Disease: Controversies in Management: Richter, B.A. (Las Vegas, USA)
Pediatric Idiopathic Hypertension (Pseudotumour Cerebri): Xu, M.W. (Sydney, N.S.W.), Li, G.S. (Philadelphia, Pa.)
Bone Age Assessment in the Workup of Children with Endocrine Disorders: Spadaro, G.L., Guevara, S. (Rome)
Obesity-Related Renal Injury in Childhood: Santos, A., Pelliccia, P., Chiarelli, F., Molas, A. (Chieti)
Insulin-Like Growth Factor-I Treatment in Primary Growth Hormone Insensitivity: Effect of Recombinant Human IGF-I (rhIGF-I) and rhIGF-I/rhIGF-Binding Protein-3 Complex: Todd, N., P. Fluck, E., Bullo, P.E. (Bern)

More information at www.karger.com/hrp
• Pay-per-view and Subscriber Access to Full Text
• Full Table of Contents
• Full Editorial Board
• Free Abstracts and Selected Articles
• Online Sample Issue
• Submission/Guidelines for Authors
• Subscription Details
• Free Alert Service
• Online Library Recommendation

Hormone Research in Paediatrics
2012 Volume 77, 78
6 issues per volume
Language: English
ISSN 0166–2818 (print)
ISSN 1663–2826 (online)

Listed in bibliographic services, including Current Contents®/Life Sciences, PubMed/MEDLINE, Biological Abstracts, Current Contents®, BMED/Biological Abstracts, Chemical Abstracts, EMBASE/Excerpta Medica

Letters to the Editor

From Animal Models to Clinical Conditions

Combining basic knowledge and clinical experience

Adipose Tissue Development

From Animal Models to Clinical Conditions

Editors
Claire Levy-Marchal
Luc Pénicaud

www.karger.com/endv

More information at www.karger.com/hrp

A qualified source of experimental data and informative clinical reports

HORMONE RESEARCH IN PÆDIATRICS
From Developmental Endocrinology to Clinical Research

Editor
P. Czernichow, Paris

Associate Editors
S. Bermon, Parma
F. Chiarelli, Chieti
W.S. Coutfeld, Auckland
M.T. Dattani, London
O. Hart, Lübeck

With some of the greatest advances from developmental endocrinology to clinical research being observed as a direct result of close collaboration between basic and clinical investigators, ‘Hormone Research in Paediatrics’ facilitates the interchange of ideas by pooling findings at all levels and from all branches side by side in a single source. Original research papers provide a background of experimental data on the pathology, cyogen, histology, biochemistry, pharmacology and regulation of hormones. This information is complemented by authoritative reports drawing on clinical experiences concerning the many diagnostic and therapeutic procedures and possibilities. In particular, such topical problems as growth, diabetes, obesity, sexual development and tumours of endocrine origin are covered in depth. As a bonus for subscribers, the journal also features frequent supplement issues.

Selected contributions
Validation of Bone Age Methods by Their Ability to Predict Adult Height: Thoburn, R.M.; Moon, J.C.; Keshel, J.; Langley, M.B. (Chicago), Jouve, A.G. (Paris), Martin, M.B. (Düsseldorf)
Pediatric Graves’ Disease: Controversies in Management: Richter, B.A. (Las Vegas, USA)
Pediatric Idiopathic Hypertension (Pseudotumour Cerebri): Xu, M.W. (Sydney, N.S.W.), Li, G.S. (Philadelphia, Pa.)
Bone Age Assessment in the Workup of Children with Endocrine Disorders: Spadaro, G.L., Guevara, S. (Rome)
Obesity-Related Renal Injury in Childhood: Santos, A., Pelliccia, P., Chiarelli, F., Molas, A. (Chieti)
Insulin-Like Growth Factor-I Treatment in Primary Growth Hormone Insensitivity: Effect of Recombinant Human IGF-I (rhIGF-I) and rhIGF-I/rhIGF-Binding Protein-3 Complex: Todd, N., P. Fluck, E., Bullo, P.E. (Bern)

More information at www.karger.com/hrp
• Pay-per-view and Subscriber Access to Full Text
• Full Table of Contents
• Full Editorial Board
• Free Abstracts and Selected Articles
• Online Sample Issue
• Submission/Guidelines for Authors
• Subscription Details
• Free Alert Service
• Online Library Recommendation

Hormone Research in Paediatrics
2012 Volume 77, 78
6 issues per volume
Language: English
ISSN 0166–2818 (print)
ISSN 1663–2826 (online)

Listed in bibliographic services, including Current Contents®/Life Sciences, PubMed/MEDLINE, Biological Abstracts, Current Contents®, BMED/Biological Abstracts, Chemical Abstracts, EMBASE/Excerpta Medica
Neonatology

Fetal and Neonatal Research


Incorporating 'Developmental Pharmacology and Therapeutics', founded by J.V. Aranda, Montreal.

Editors-in-Chief

H.L. Halliday, Belfast
C.P. Speer, Würzburg

Editorial Board

S. Andersson, Helsinki
E. Bancalari, Miami, Fla.
J. Bhatia, Augusta, Ga.
G. Buonocore, Siena
W. Carlo, Birmingham, Ala.
I. Choonara, Derby
T. Curstedt, Stockholm
C. Dani, Florence
B. Darlow, Christchurch
M. Fujimura, Osaka
M. Hallman, Oulu
W.W. Hay, Jr., Aurora, Colo.
S.E. Juul, Seattle, Wash.
M. Kaplan, Jerusalem
B. Kramer, Maastricht
R.J. Martin, Cleveland, Ohio
C.J. Morley, Cambridge
J. Neu, Gainesville, Fla.
P.C. Ng, Hong Kong

M.W. Obladen, Berlin
A.G.S. Philip, Sebastopol, Calif.
M. Post, Toronto
E. Saliba, Tours
O.D. Saugstad, Oslo
M.S. Schimmel, Jerusalem
M.P. Sherman, Columbia, Mo.
E.S. Shinwell, Rehovot
K. Simmer, Perth, W.A.
J. Smith, Tygerberg
B. Sun, Shanghai
N. Vain, Buenos Aires
F. van Bel, Utrecht
J.N. van den Anker, Washington, D.C.
M. Vento Torres, Valencia
M. Weindling, Liverpool
J.A. Widness, Iowa City, Iowa
Guidelines for Authors

Aims and Scope
This highly respected and frequently cited journal is a prime source of information in the area of fetal and neonatal medicine. Original Papers present research on all aspects of neonatology, fetal medicine and developmental biology. These papers encompass both basic science and clinical research including randomised trials, observational studies and epidemiology. Basic science research covers molecular biology, molecular genetics, physiology, biochemistry and pharmacology in fetal and neonatal life. Papers reporting results of animal studies should be based upon hypotheses that relate to developmental processes or disorders in the human fetus or neonate.

Submission
Manuscripts written in English should be submitted using the online submission website at: www.karger.com/neo or as e-mail attachment (the preferred word-processing package is MS-Word) to the Editorial Office: neo@karger.ch

S. Karger AG
Editorial Office 'Neonatology'
P.O. Box
CH–4009 Basel (Switzerland)

All manuscripts must be accompanied by a cover letter and a statement with all authors' signatures (handwritten) saying that they agree with the publication of the paper. Names, postal and e-mail addresses of four international experts in the appropriate area of research should accompany each manuscript. Selected scientists(s) will be invited to act as referee(s). Referees suggested should not be from the same institution as the author and should have expert knowledge of the subject.

Manuscripts may be submitted to the following sections:
- Editorials
- Reviews
- Original Papers
- Consensus Statements
- Short Communications
- Novel Insights from Clinical Practice
- Commentaries
- Research Briefings
- Sources of Neonatal Medicine
- Letters to the Editor

Reviews
Most reviews are submitted upon invitation. However, the editors are open to unsolicited reviews. Authors planning such a review are requested to contact the Editorial Office with a one page outline of the intended review. All reviews are subject to peer review. Systematic reviews should be reported using the format of the Cochrane Neonatal Review Group (www.neonatal.cochrane.org/ welcome).

Original Papers
Original articles should not exceed a printed length of two pages, i.e. generally not more than 1,400 words of text accompanied by 2 figures or tables and from 5 to 10 references.

Novel Insights from Clinical Practice (formerly Case Reports)
The publication space available for case reports is very limited. The journal only considers case reports with significant new insights or with an extremely unusual and memorable course. Highlighted boxes containing one or two bullet points on 'Established Facts' (what is already known) and 'Novel Insights' (what new information has been gained) are required and should be placed on the first page of the report. These should be selected so as to reinforce the novelty of the clinical observation. The manuscript should be presented with an abstract (unstructured, max. 150 words), follow by introduction, case report and discussion. Maximum 2 figures or 1 tables or 1 figure and 1 table.

Conditions
All manuscripts are subject to editorial review. Manuscripts are received with the explicit understanding that they are not under simultaneous consideration by any other publication. Submission of an article for publication implies the transfer of the copyright from the author to the publisher upon acceptance. Accepted papers become the permanent property of 'Neonatology' and may not be reproduced by any means, in whole or in part, without the written consent of the publisher. It is the author's responsibility to obtain permission to reproduce illustrations, tables, etc. from other publications.

Conflict of Interest: All forms of support, including that from drug companies, and any potential source of conflict of interest should be acknowledged in the cover letter to the editor when applicable. The statement will be printed at the end of the article.

Ethics: Published research must comply with the guidelines for human studies and animal welfare regulations. Authors should state that subjects have given their informed consent and that the study protocol has been approved by the institution's committee on human research. Further, they should also state that animal experiments conform to institutional standards.

Arrangement
Title page: The first page of each paper should indicate the title, the authors' names, the institute where the work was conducted, and a short title for use as running head. The title should be continuous throughout the entire manuscript (i.e., do not begin numbering from 1 again at the top of each page).

Abstract: Each paper needs an abstract of up to 250 words. It should be structured as follows:
- Background: What is the major topic that prompted the study?
- Objectives: What is the purpose of the study?
- Methods: How was the study done?
- Results: Most important findings
- Conclusions: Most important conclusions

Footnotes: Avoid footnotes.

Tables and illustrations: Tables and illustrations (both numbered in Arabic numerals) should be prepared on separate pages. Tables require a heading and figure legends should be supplied on a separate page. For technical reasons, figures with a screen background should not be submitted. When possible, group several illustrations on one block for reproduction (max. size 180 × 223 mm) or provide crop marks. Electronically submitted b/w half-tone and color illustrations must have a final resolution of 300 dpi after scaling, line drawings one of 800–1200 dpi. Figure files must not be embedded in a document file but submitted separately.

Color illustrations: Online edition: Color illustrations are reproduced free of charge. In the print version, the illustrations are reproduced in black and white. Please avoid referring to the colors in the text and figure legends.

Print edition: Up to 6 color illustrations per page can be integrated with the text. Additional color illustrations can be ordered at CHF 800.– per page.

References: In the text identify references by Arabic numerals [in square brackets]. Material submitted for publication but not yet accepted should be noted as 'unpublished data' and not be included in the reference list. The list of references should include only those publications which are cited in the text. Do not alphabetize; number references in the order in which they are first mentioned in the text. The surnames of the authors followed by initials should be given. There should be no punctuation other than a comma to separate the authors. Preferably, please cite all authors. Abbreviate journal names according to the Index Medicus system. Also see International Committee of Medical Journal Editors: Uniform requirements for manuscripts submitted to biomedical journals (www.icmje.org).

Examples
(b) Papers published only with DOI numbers: AllFah K, Anabrees J, Basler D: Probiosis reduces the risk of necrotizing enterocolitis in preterm infants: a meta-analysis. Neonatology DOI: 10.1159/000235684.

SI Units
SI units should be used. Listings of SI units may be found in the following publications: Lundberg GD, Iversen C, Raudalescu G: Now read this: the SI units are here. JAMA 1986;255:2329–2339.


Digital Object Identifier (DOI)
S. Karger Publishers supports DOIs as unique identifiers for articles. A DOI number will be printed on the title page of each article. DOIs can be useful in the future for identifying and citing articles published online without volume or issue information. More information can be found at www.doi.org.

Supplementary Material
Supplementary material is restricted to additional data that are not necessary for the scientific integrity and conclusions of the paper. Please note that all supplementary files will undergo editorial review and should be submitted together with the original manuscript. The Editors reserve the right to limit the scope and length of the supplementary material. Supplementary material must meet production quality standards for Web publication without the need for any modification or editing. In general, all files should be supplied separately and named clearly. Acceptable files and formats are: Word or PDF files, Excel spreadsheets (only if the data cannot be converted properly to a PDF file), and videos files (.mov, .avi, .mpeg).

Author’s ChoiceTM
Karger’s Author’s ChoiceTM service broadens the reach of your article and gives all users worldwide free and full access for reading, downloading and printing at www.karger.com. The option is available for a one-time fee of CHF 3000.–, which is a permissible cost in grant allocation. More information can be found at www.karger.com/authors_choice.

NIH-Funded Research
The U.S. National Institutes of Health (NIH) mandates under the NIH Public Access Policy that final, peer-reviewed manuscripts appear in its digital database within 12 months of the official publication date. As a service to authors, Karger submits the final version of your article on your behalf to PubMed Central. For those selecting our premium Author’s ChoiceTM service, we will send your article immediately upon publishing, accelerating the accessibility of your work without the usual embargo. More details on NIH’s Public Access Policy are available at http://publicaccess.nih.gov/policy.htm

Self-Archiving
Karger permits authors to archive their pre-prints (i.e. pre-refereeing) or post-prints (i.e. final draft post-refereeing) on their personal or institution’s servers, provided the following conditions are met: Articles may not be used for commercial purposes, must be linked to the publisher’s version, and must acknowledge the publisher’s copyright. Authors selecting Karger’s Author’s ChoiceTM feature, however, are also permitted to archive the final, published version of their article, which includes copy-editing and design improvements as well as citation links.

Page Charges
A charge of CHF 60.– per page (except for invited reviews which are free) will be levied for the first 3 printed pages of an article. Each additional complete or partial page will be charged to the author at CHF 325.–. 3 printed pages are equal to approx. 9 manuscript pages (including tables, illustrations and references).

Proofs
Proofs are sent to the corresponding author and should be returned with the least possible delay. Alterations other than the correction of printer’s errors are charged to the author.

Reprints
Order form and price list is sent with the pdf proofs. Orders submitted after the issue is printed are subject to considerably higher prices.

“A collection of extraordinary essays”

GOTTFRIED SCHATZ
A MATTER OF WONDER
What Biology Reveals about Us, Our World, and Our Dreams

Where do we come from? Is our destiny determined by the genes we inherit? In this book Gottfried Schatz, the world-renowned biochemist and co-discoverer of mitochondrial DNA, gives lucid – albeit often surprising – answers to universal questions and takes the reader on a fascinating journey of discovery across the boundaries of scientific disciplines. With passion and a keen sense of wonder he draws on philosophy, cultural history and art to formulate his reflections on the mysteries of life. His essays will appeal not only to scientists but to all inquisitive minds, regardless of educational and professional background.

G. Schatz (Basel)
A Matter of Wonder
What Biology Reveals about Us, Our World, and Our Dreams
Translated by A. Shields
XII + 190 p., 2 color fig., hard cover, 2011
CHF 29.– / EUR 21.50 / USD 29.00

More information and sample essays at
www.karger.com/schatz

S. Karger AG, P.O. Box
CH–4009 Basel
(Switzerland)
Fax +41 61 306 12 34
E-Mail orders@karger.ch
www.karger.com

© 2012 S. Karger AG, Basel
The Guidelines for Authors are available at:
www.karger.com/neo_Guidelines

www.karger.com
General Information

Subscription Rates: Subscriptions run for a full calendar year. Prices are given per year.

- Personal subscription:
  - Print or Online: CHF 721.–, EUR 577.–, USD 700.00
  - Postage and handling (added to print and print+online): CHF 54.40 Europe, CHF 80.– Overseas
- Print or Online:
  - CHF 2884.–, EUR 2308.–, USD 2800.00
  - Postage and handling (added to print and print+online): CHF 68.– Europe, CHF 100.– Overseas
  - EUR 52.–, USD 64.00

Institutional subscription:
- Print or Online:
  - CHF 3172.–, EUR 2538.–, USD 3080.00
  - Airmail surcharge: CHF 68.– / USD 74.00

Discount subscription prices:
- Personal or internal use of specific clients, provided that the stated fee is paid per copy directly to S. Karger AG, P.O. Box CH–4009 Basel (Switzerland).
- Posters, data, or electronic back volumes can be obtained from Copyright Clearance Center Inc., 222 Rosewood Drive Danvers, MA 01923 (USA)

Photocopying:
- It has been registered with the Copyright Clearance Center (CCC), as indicated by the code appearing on the first page of each article. For readers in the US, this code signals consent for copying of articles for personal or internal use, or for the personal or internal use of specific clients, provided that the stated fee is paid per copy directly to Copyright Clearance Center Inc.

Change of Address:
Both old and new address should be sent to the subscription source.
36th Annual International Conference

“Miami Neonatology 2012”

Workshop:

“Advances in Neonatal Respiratory Care”

October 31 – November 3, 2012
Fontainebleau, Miami Beach

Speakers

Olaf Bodamer, M.D. PhD FACMG (Miami, FL)
Nicholas J. Evans, M.D. (Sydney Australia)
Petra Huppi, M.D. (Geneva Switzerland)
Paolo Marzoni, M.D. (Torino, Italy)
Roberto Romero, M.D. (Detroit, MI)
Juan E. Sola, M.D. (Miami, FL)
Robin H. Steinhorn, M.D. (Chicago, IL)
Benjamin Stenson, M.D. (Edinburgh, United Kingdom)
Rose M. Viscardi, M.D. (College Park, MD)
Ekhard Ziegler, M.D. (Iowa City, IA)

Topics

• PDA, systemic blood flow and circulatory support
• Use of point care ultrasound in the NICU
• Lacteal in newborns and prevention of neonatal sepsis
• New strategies for the management of neonatal candida sepsis
• Nutritional strategies to decrease the burden of neonatal infections
• Intrauterine infection/inflammation in preterm labor and neonatal injury
• Cervical ultrasound and vaginal progesterone to prevent preterm delivery
• Radical thoughts about oxygen
• Pulmonary vascular disease and prematurity
• Practical issues of targeting oxygen in the delivery room and neonatal ICU
• Diagnosis of sepsis using microarray
• Developmental outcomes of the SUPPORT trial
• Global health aspects of neurodevelopmental disabilities
• Ureaplasma colonization and respiratory outcome
• Ventilator associated pneumonia

http://pediatrics.med.miami.edu/neonatology/international-neonatal-conference

20th Annual International Neonatal Conference

14 to 16 June 2012
Wynyard Rooms, Wynyard Park, Billingham
Teesside, TS22 5TB

Pre-conference “Advanced Ventilatory Workshop” - 13 June 2012

Annual Neonatal Conference - 14 to 16 June 2012 (3 days)

Topics include:


Registration Contact

Conferences and Courses Department, Academic Centre, The James Cook University Hospital, Marton Road, Middlesbrough, TS4 3BW
Telephone 01642 282534 / 01642 282825, Fax 01642 282535, Email nicky.skippon@stees.nhs.uk

APPLY ONLINE - WWW.NEONATALCONFERENCE.CO.UK
39th ANNUAL MEETING OF THE
Fetal and Neonatal Physiological
SOCIETY

8-11 JULY 2012 - UTRECHT, THE NETHERLANDS

ORGANISING COMMITTEE

Jan Derks, Joepe Kaandorp, Edu Mulder, Deodata Tijsseling, Gerard Visser

Please note these dates!
8-11 July 2012

University Medical Center
Utrecht

WWW.FNPS2012.NL

Conference Secretariat: Klinkhamer Conference Management P.O. Box 1308 6201 BH Maastricht The Netherlands T +31 (0)43-36 27 008 E info@fnps2012.nl
Contents

See the journal website for contents
CIPP XI 11th International Congress on Pediatric Pulmonology

Bangkok, Thailand
June 30 - July 2, 2012

Deadlines:
Abstract submission: December 30, 2011
Early bird registration: March 1, 2012

www.cipp-meeting.org  cipp@cipp-meeting.org

Medi@xa
27 Rue Masséna, 06000 Nice, France
tel: 0033 4 97 03 85 97
fax: 0033 4 97 03 85 98
Both parents and offspring are susceptible to adverse environmental conditions that alter their normal brain development and adaptations during reproduction, increasing their risk of mental problems in the short and long term. Pregnancy stress and anxiety alter the cognitive performance, memory, and behavior of mothers. Resulting in suboptimal maternal hormonal signals and inadequate care, they impact directly and indirectly on the developing baby in utero and in the neonatal stage.

This special issue of Neuroendocrinology is a collection of timely review articles from experts in the field of Mental Health Programming presented at the 'Parental Brain' Conference in Edinburgh in September 2010. A variety of mental health topics ranging from the neonatal to the juvenile and parental brain are discussed in detail.

The insight provided here from in-depth research into brain mechanisms underlying altered mental health marks the recent realization that mental health is susceptible to adverse programming from an early age and that real harm can be passed on inadvertently from generation to generation. Therefore 'The Parental Brain' offers valuable reading for scientists and clinicians interested in the impact of environmental conditions on mental health and how parental health contributes to long-term mental health in offspring.
Adipose Tissue Development
From Animal Models to Clinical Conditions

Editors
Claire Levy-Marchal
Luc Pénicaud

Combining basic knowledge and clinical experience

Nowadays, adipose tissue is not only regarded as an organ of storage related to fuel metabolism but also as an endocrine organ involved in the regulation of insulin sensitivity, lipids and energy metabolism.

These proceedings cover the nervous regulation of both white and brown adipose tissue mass. Different physiological parameters such as metabolism (lipolysis and thermogenesis) and secretory activity (leptin and other adipokines) are reviewed. The plasticity of adipose tissue (proliferation, differentiation and apoptosis) showing the presence of a neural feedback loop between adipose tissue and the brain, which plays a major role in the regulation of energy homeostasis, is discussed.

Merging basic knowledge and various clinical conditions, this thorough review is of great interest to both scientists and physicians, in particular, pediatrists, interested in obesity, endocrinology and nutrition.

Contents
Preface: Levy-Marchal, C.
Human Lipodystrophies: Genetic and Acquired Diseases of Adipose Tissue: Capron, J.; Mogeri, J.; Cossu-Dekterer, M.; Lagattu, C.; Amstutz, B.; Bertrand, V.; Lebret, C.; Restelli, J.-F.; Bagliero, C.
The Emergence of Adipocytes: Lahortigue, P.; Castella, L.
Adipose Tissue and the Reproductive Axis: Biological Aspects: Nowoum, G.; Zobic, C.R.
Unraveling the Obesity and Breast Cancer Links: A Role for Cancer-Associated Adipocytokines: Dufour, B.; Rechel, L.; Acenauer, G.; Velut, P.; Müller, C.
Early Determinants of Obesity: Ong K.K.
Metabolic Syndrome in Childhood – Causes and Effects: Weiss, K.
Pathophysiology of Insulin Resistance in Small for Gestational Age Subjects: A Role for Adipose Tissue?: Beltrand, J.; Mines, T.; Levy-Marchal, C.
The Neural Feedback Loop between the Brain and Adipose Tissue: Pénicaud, L.

Author Index
Subject Index
Molecular mechanisms, clinical manifestations, and new treatments

Monogenic Hyperinsulinemic Hypoglycemia Disorders

Editors
Charles A. Stanley
Diva D. De León

Contents
Preface: Stanley, C.A.; De León, D.D.
Historical Perspective on the Genetic Forms of Congenital Hyperinsulinism: Stanley, C.A.; Matschinsky, F.M.
Biochemistry and Physiology of the ATP-Sensitive Potassium Channel: Aten, L.J.; Adzick, N.S.
Pathophysiology of Diffuse ATP-Sensitive Potassium Channel Hypersensitivity: De León, D.D.; Stanley, C.A.
Molecular Defects of ATP-Sensitive Potassium Channels in Congenital Hyperinsulinism: Aten, L.J.; Shyng, S.-L.; Bushman, J.D.; Pratt, E.B.; Zhou, Q.; Stanley, C.A.
Genetic Loci Involved in Congenital Hyperinsulinism: Molven, A.; Helgeland, G.; Servoin, J.-B.; Njølstad, P.R.
Glutamine Dehydrogenase: Structure, Regulation, and Its Role in Insulin Homeostasis: Smith, T.J.; Sayed, S.; Matschinsky, F.M.; Stanley, C.A.
Clinical Manifestations Due to Mutations of Glutamate Dehydrogenase: The Hypoglycemia/Hyperammonemia Syndrome: Carobbio, S.; Sayed, S.; Matschinsky, F.M.; Stanley, C.A.
Amino Acid-Stimulated Insulin Secretion: The Role of the Glutaminol-Glutamate-Alpha-Ketoglutarate Rate Axis: L.C.; Matschinsky, F.M.; Stanley, C.A.
Molecular Genetics and Pathophysiology of Congenital Hyperinsulinism Caused by Short Chain 3-Hydroxycyclo-Carboxylate Dehydrogenase Deficiency: Malmberg, S.; Helgeland, G.
Pancreatic Histopathology of Hyperinsulinism: Verkarre, V.; Rossignol, S.; Aigrain, Y.; de Lonlay, P.
Molecular Mechanisms and Clinical Pathophysiology of Focal ATP-Sensitive Potassium Channel Hypersensitivity: Said, S.; Ruther, E.D.
Localisation of a Focal Loss of Congenital Hyperinsulinism in Imaging and Surgery: Jarets, L.; Adibekian, A.N.
Role of Incretin Hormones in Hyperinsulinemic Disorders: De León, D.D.
Glutamine Dehydrogenase: Structure, Regulation, and Its Role in Insulin Homeostasis: Smith, T.J.; Sayed, S.; Matschinsky, F.M.; Stanley, C.A.
Hyperinsulinism Due to Mutations of Uncoupling Protein 2: Bar Mat-Ganzarain, M.; de Lonlay, P.; Rischin, D.
Exercise-Induced Hyperinsulinism: A Failure of Monocarboxylate Transporter 1 Expression Silencing: Oostrum, I.; Moes, B.; Meijer, P.; van Vugt, M.; Matschinsky, F.M.; van der Velden, J.; Rood, C.; Hassink, A.

Author Index
Subject Index

Systematic Review and Meta-Analysis

183 Surfactant Lavage Therapy for Meconium Aspiration Syndrome: A Systematic Review and Meta-Analysis

Sources of Neonatal Medicine

159 Thrush – Nightmare of the Foundling Hospitals
Obal-B, M. (Berlin)

Original Papers

166 Noninvasive Assessment of the Early Transitional Circulation in Healthy Term Infants
Popat, H.; Khidrow, M. (Sydney, N.S.W.)

172 Recognition, Diagnosis and Treatment of Meconium Obstruction in Extremely Low Birth Weight Infants

179 First Day of Life Reference Values for Pleth Variability Index in Spontaneously Breathing Term Newborns
Latini, G. (Brindisi/Pisa); Dipuola, L. (Pisa); De Felice, C. (Nerina)

192 Differential Hemodynamic Effects of Levosimendan in a Porcine Model of Neonatal Hypoxia-Reoxygenation
Bae, C.-W. (Seoul); Cheung, P.-Y. (Edmonton, Alta.); Biggan, D. (Edmonton, Alta.); Vittorio, M. (Valencia); Cheng, P.-Y. (Edmonton, Alta.)

Thewissen, L.; Allegaert, K. (Leuven)

217 Postnatal Rosiglitazone Administration to Neonatal Rat Pups Does Not Alter the Young Adult Metabolic Phenotype
Truong, N.C.; Abbassi, A.; Sakurai, R.; Lee, W.N.; Pordy, J.S.; Rehnau, V.K. (Torrance, Calif.)

225 Is Octreotide Treatment Useful in Patients with Congenital Chylothorax?
Harvey, M.; Moong, C.F.; Antebi, T.A.J. (Nijmegen)

232 Efficacy and Safety of a Tight Glucose Control Protocol in Critically Ill Term Neonates
Verbruggen, S.C.; Landzart, J.-I.; Reins, I.K.M. (Rotterdam); van Goudoever, J.B. (Rotterdam/Amsterdam); Joosten, K.F.M. (Rotterdam)

Novel Insights from Clinical Practice

201 Levosimendan in Two Neonates with Ischemic Heart Failure and Pulmonary Hypertension
De Carolis, M.P.; Piazza, M.; Bersani, I.; Pardo, M.; Stival, E.; Tempera, A.; Romagoli, C.; Conti, G.; De Rosa, G. (Rome)

Commentary

210 Cognitive and Neurological Outcome at the Age of 3–8 Years of Preterm Infants with Prolonged Ventricular Dilation Requiring Neurosurgical Intervention