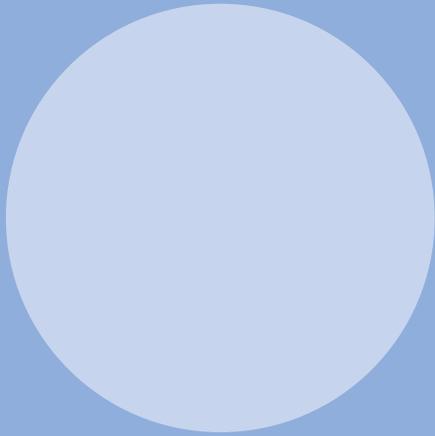


Cardiology in the Young



Submit your paper online
<http://mc.manuscriptcentral.com/cty>

CAMBRIDGE
UNIVERSITY PRESS

INVOS

REVEALING BRIGHTER FUTURES

SYSTEM



Someday I want to be a
doctor, not a patient.

His heart surgery seemed to go smoothly. But will kindergarten?

Brain dysoxygenation. Shock. Renal failure. Congenital heart patients face numerous ischemic threats, but the INVOS® System can help keep them safe. Oxygenation data from up to four specific sites helps you assess perfusion distribution and oxygen saturation in real time, allowing for early detection and rapid response. It also shows immediate patient responses to interventions so you can better judge their impact and efficacy. So let the INVOS System help these kids navigate through the OR and PICU... and perhaps even kindergarten too¹⁻³.



INVOS CEREBRAL/SOMATIC
OXIMETER
REFLECTING THE COLOR OF LIFE®

1. Hoffman et al, *Anesthesiology* 2008; 109 A7 2. Dent et al, *J Thorac Cardiovasc Surg.* 2006 Jan;131(1):190-7
3. Andropoulos et al, *Anesthesiology* 2007; 107: A209

SOMANETICS®

www.somanetics.com | 800.359.7662

Cardiology in the Young

journals.cambridge.org/CTY

Editor-in-Chief

Dr Edward J. Baker

Department of Congenital Heart Disease, Evelina Children's Hospital,
Guy's & St. Thomas' Hospital NHS Trust, Lambeth Palace Road, London, SE1 7EH, UK

Emeritus Founding Editor

Professor Robert H. Anderson

Cardiac Unit, Institute of Child Health, University College London,
30 Guilford Street, London WC1N 1EH, UK

International Editors

G. William Henry, Chapel Hill, NC
Hiromi Kurosawa, Tokyo

Supplements Editor Gil Wernovsky, Philadelphia, PA

Images Editor Roxane McKay, Memphis, TN

Founding Editors

Anton E. Becker, Amsterdam; Giancarlo Crupi, Bergamo; Arthur Garson Jr,
Charlottesville, VA; Fernando Lucchese, Porto Alegre; Lucio Parenzan,
Bergamo; the late Atsuyoshi Takao, Tokyo; Michael Tynan, London

International Editorial Board

Lindsey D. Allan (London)
Maurice Beghetti (Geneva)
Lee N. Benson (Toronto)
Per G. Bjørnstad (Oslo)
Anthony C. Chang (Orange, CA)
Tjark Ebels (Groningen)
Timothy Feltes (Columbus, OH)
J. William Gaynor (Philadelphia, PA)
Allan Goldman (London)
Willem A. Helbing (Rotterdam)
John Hess (Munich)
Jeffrey P. Jacobs (St Petersburg, FL)
Eero Jokinen (Helsinki)
Tom Karl (Brisbane)
Doff B. McElhinney (Boston, MA)

Luc Mertens (Leuven)
Cleonice de C. Mota (Belo Horizonte)
Jane Newburger (Boston, MA)
Edgardo E. Ortiz (Quezon City)
Daniel Penny (Melbourne)
Andrew N. Redington (Toronto)
Girish S. Shirali (Charleston, SC)
Norman H. Silverman (Palo Alto, CA)
Giovanni Stellin (Padova)
András Szatmári (Budapest)
Hideki Uemura (London)
Steven A. Webber (Pittsburgh, PA)
James L. Wilkinson (Melbourne)
Shi-Joon Yoo (Toronto)

Cardiology in the Young is indexed and abstracted in Index Medicus/MEDLINE; Current Contents/Clinical Medicine; Research Alert; Sci Search; EMBASE/Excerpta Medica

Commissioning Editor: Dan Edwards
Production Editor: Nicki Marshall

Design and Production: Macmillan India Limited, Bangalore
Printed & Bound: Latimer Trend, Plymouth

Published by Cambridge University Press (Journals), Edinburgh Building, Shaftesbury Road, Cambridge CB2 8RU, UK
Tel: +44 (0)1223 326491; Fax: +44 (0)1223 325802; E-mail: dedwards@cambridge.org

Cambridge Journals Online

For further information about this journal please
go to the journal website at:
journals.cambridge.org/CTY



Mixed Sources
Product group from well-managed
forests and other controlled sources
www.fsc.org Cert no. SGS-COC-005493
© 1996 Forest Stewardship Council

CAMBRIDGE
UNIVERSITY PRESS

Cardiology in the Young

COPYRIGHT © 2009 CAMBRIDGE UNIVERSITY PRESS
ISSN 1047-9511

Table of Contents October 2009

FROM THE EDITOR-IN-CHIEF	427
EDITORIAL	
At what level of blood pressure should hypertension be defined in children? <i>Manish D. Sinha, Christopher J.D. Reid</i>	428
CONTINUING MEDICAL EDUCATION	
Measurement, interpretation and use of haemodynamic parameters in pulmonary hypertension associated with congenital cardiac disease <i>Antonio Augusto Lopes, Patrick W. O'Leary</i>	431
ORIGINAL ARTICLES	
Measurements of adiposity and high blood pressure among children and adolescents living in Belo Horizonte <i>Robespierre C. Ribeiro, Joel A. Lamounier, Reynaldo G. Oliveira, Isabela M. Bensenor, Paulo A. Lotufo</i>	436
Prevalence of increased nuchal translucency in fetuses with congenital cardiac disease and a normal karyotype <i>Melanie Vogel, Gurleen K. Sharland, Doff B. McElbinney, Vita Zidere, John M. Simpson, Owen I. Miller, Lindsey D. Allan</i>	441
The evolution of the pulmonary arterial sling syndrome, with particular reference to the need for reoperations because of untreated tracheal stenosis <i>Tsvetomir S. Loukanov, Christian Sebening, Wolfgang Springer, Siegfried Hagl, Matthias Karck, Mathias Gorenflo</i>	446
Mid-term results, and therapeutic management, for patients suffering hypertension after surgical repair of aortic coarctation <i>Ugo Giordano, Barbara Cifra, Salvatore Giannico, Attilio Turchetta, Armando Calzolari</i>	451
Profiles of levels of lipids and dyslipidaemia in children from Beijing, China <i>Yan Hui, Mi Jie, Liu Ying, Jin Hongfang, Chen Li, Tang Chaoshu, Du Junbao</i>	456
Non-invasive measurement of the response of right ventricular pressure to exercise, and its relation to aerobic capacity <i>Thomas Möller, Kari Peersen, Eirik Pettersen, Erik Thaulow, Henrik Holmström, Per Morten Fredriksen</i>	465
Noninvasive methods of accurately diagnosing in children anomalous origin of the left coronary artery from the pulmonary trunk <i>Kuang-Jen Chien, Ta-Cheng Huang, Kai-Sheng Hsieh, Chu-Chuan Lin, Ken-Pen Weng, Jun-Yen Pan, Cheng-Liang Lee</i>	474
Genetic screening of 104 patients with congenitally malformed hearts revealed a fresh mutation of <i>GATA4</i> in those with atrial septal defects <i>Haruka Hamanoue, Sri Endah Rahayuningsih, Yuya Hirabara, Junko Itob, Utako Yokoyama, Takeshi Mizuguchi, Hirotomo Saitou, Noriko Miyake, Fumiki Hirabara, Naomichi Matsumoto</i>	482
Diagnosis, clinical features, management, and post-natal follow-up of fetal tachycardias <i>Rajka Lulić Jurjević, Tomaž Podnar, Samo Vesel</i>	486
Reliability of echocardiographic measurements of left cardiac structures in healthy children <i>Miranda J.J. Geelboed, Sonja P.E. Snijders, Veronica E. Kleyburg-Linkers, Eric A.P. Steegers, Lennie van Osch-Gevers, Vincent W.V. Jaddoe</i>	494

Table of Contents *continued*

Coenzyme Q10 improves diastolic function in children with idiopathic dilated cardiomyopathy <i>Armen Kocharian, Reza Shabaniyan, Mohammad Rafiei-Khorgami, Abdolrazagh Kiani, Giv Heidari-Bateni</i>	501
Perceptions of caregivers following percutaneous endoscopic gastrostomy in children with congenitally malformed hearts <i>Ramesh Srinivasan, Catherine O'Neill, Wendy Blumenow, A. Mark Dalzell</i>	507
Causes of death and cardiovascular complications in adolescents and adults with congenitally malformed hearts: an autopsy study of 102 cases <i>Vera Demarchi Aiello, Maria Angélica Binotto, Lea Maria Demarchi, Antonio Augusto Lopes, Miguel Barbero Marcial</i>	511
IMAGES IN CONGENITAL CARDIAC DISEASE	
Radiofrequency perforation of the pulmonary valve in an adult with tetralogy of Fallot and pulmonary atresia <i>Arif Anis Khan, Michael J. Mullen, Alan G. Magee</i>	517
BRIEF REPORTS	
Single-stage correction, with extensive pulmonary arterial reconstruction, of a pulmonary arterial sling associated with tetralogy of Fallot <i>Onur S. Goksel, Emin Tireli, Ahmet Çelebi</i>	519
Pulmonary atresia with intact ventricular septum associated with aortic coarctation <i>Arjamand Shauq, Gordon Gladman, Edmund J. Ladusans</i>	522
Complete congenital obstruction of a scimitar vein in an infant with severe respiratory failure <i>Alexandra Bauch, Kyong-Jin Lee, Shi-Joon Yoo</i>	524
Totally anomalous pulmonary venous connection to superior caval vein after an intracytoplasmic injection of sperm <i>Javier Pérez-Lescure Picarzo, David Crespo Marcos</i>	527
An intrathoracic arteriovenous malformation discovered as an extremely uncommon reason of neonatal congestive cardiac failure <i>Sigrun R. Hofmann, Matthias Weise, Katharina I. Nitzsche</i>	530
Catheter closure of an atrial septal defect in anatomically corrected malposition with left juxtaposition of atrial appendages <i>Kothandam Sivakumar</i>	534
A patient with aneurysms of both aortic coronary sinuses of Valsalva obstructing both ventricular outflow tracts <i>Isbwarappa B. Vijayalakshmi, Nijagal S. Devananda, Narasimban Chitra</i>	537
Defects in long chain fatty acid oxidation presenting as severe cardiomyopathy and cardiogenic shock in infancy <i>Narendra R. Dereddy, David Kronn, Usba Krishnan</i>	540
ERRATUM	543

This journal issue has been printed on FSC-certified paper and cover board. FSC is an independent, non-governmental, not-for-profit organization established to promote the responsible management of the world's forests. Please see www.fsc.org for information

Cardiology in the Young © 2009 Cambridge University Press, ISSN 1047-9511 is published bi-monthly

Subscription information: Volume 19 (6 issues) will appear in 2009. Correspondence concerning subscriptions should be addressed to: Journals Customer Services, Cambridge University Press, The Edinburgh Building, Cambridge CB2 8RU, UK. Tel: +44 (0)1223 326070; Fax: +44 (0)1223 325150; E-mail enquiries: journals_subscriptions@cambridge.org; Web: www.cambridge.org; Cambridge Journals Online: www.journals.cambridge.org. The subscription rate for 2009 Volume 19 (6 issues) is: Individual rate £290 or US\$475; Institutional rate £565 or US\$930. All prices inclusive of postage. Cheques should be made payable to Cambridge University Press. Copyright: The submission of a manuscript implies the following: (a) that the work described has not been published before, except in the form of an abstract or as part of a published lecture, review, or thesis; (b) that it is not under consideration for publication elsewhere; (c) that all co-authors approve its publication; (d) that its publication is approved by the responsible authorities at the institute where the work has been carried out; (e) that when the manuscript is accepted for publication, the authors agree to the automatic transfer of the copyright to the publisher; (f) that the manuscript will not be published elsewhere in any language without the consent of the copyright holders, and (g) that written permission of the copyright holder is obtained by the authors for material used from other copyrighted sources. All articles published in this journal are protected by copyright, which covers the exclusive rights to reproduce and distribute the article (e.g., as offprints), as well as all translation rights. No material published in this journal may be reproduced photographically or stored on microfilm, in video disks, etc., without first obtaining written permission from the publishers. The use of general descriptive names, trade names, trademarks, etc., in this publication, even if not specifically identified, does not imply that these names are not protected by the relevant laws and regulations. While the advice and information in this journal are believed to be true and accurate at the date of its going to press, neither the authors, the editors, nor the publisher can accept any legal responsibility for any errors or omissions that may be made. The publisher makes no warranty, express or implied, with respect to the material contained herein. Cambridge University Press publishes advertisements in this journal while relying on the responsibility of the advertiser to comply with all legal requirements relating to the marketing and sale of products or services advertised. Cambridge University Press and the editors are not responsible for claims made in the advertisements published in the journal. The appearance of advertisements in Cambridge University Press publications does not constitute endorsement, implied or intended, of the product advertised or the claims made for it by the advertiser. Photocopies may not be made for personal or in-house use beyond the limitations stipulated under US Copyright Law. Advertisements: For advertisement rates and information on advertisement copy contact Cardiology in the Young, Advertising Dept., Cambridge University Press (Journals), Edinburgh Building, Shaftesbury Road, Cambridge CB2 8RU, UK. Tel: +44 (0)1223 325083; Fax: +44 (0)1223 325801; E-mail: ad_sales@cambridge.org. Change of address: Please notify the Subscription Department. Send new address and postal code, and the address label from a current issue to Journals Customer Services, Cambridge University Press, FREEPOST CB27, The Edinburgh Building, Cambridge CB2 8BR, UK. Tel: +44 (0)1223 326070; Fax: +44 (0)1223 325150; E-mail enquiries: journals_subscriptions@cambridge.org; Web: www.cambridge.org; Cambridge Journals Online: www.journals.cambridge.org. Printing: Cardiology in the Young (ISSN 1047-9511) is printed bi-monthly in 2009 by Latimer Trend, Plymouth.

CARDIOLOGY IN THE YOUNG

Submission to *Cardiology in the Young* is exclusively via the web-based peer-review system, CTY Manuscript Central.

Online submission enables rapid review and allows online manuscript tracking.

We invite all authors to submit online any NEW MANUSCRIPTS that are to be considered for publication in *Cardiology in the Young*.

Please use the following URL: <http://mc.manuscriptcentral.com/cty>

Editorial policies

Cardiology in the Young is devoted to cardiovascular issues affecting the young and the older patient with the sequels of cardiac disease acquired in childhood. Submission of both basic research and clinical papers is encouraged. Articles on fundamental principles will also be considered for publication. Reviews on recent developments are welcome. The Journal serves the interest of all professionals concerned with these topics. By design, the Journal is international and multidisciplinary in its approach, and the members of the Editorial Board take an active role in the Journal's mission. Prospective authors are encouraged to consult with the editors and members of the Editorial Board with any inquiries. The editors encourage the submission of articles from developing countries.

Articles should be concerned with original research not published previously and not being considered for publication elsewhere. Submission of a manuscript to the Journal gives the publisher the right to publish that paper if it is accepted, and the copyright of the manuscript becomes property of the publisher. Manuscripts may be edited to improve clarity and expression.

Authors must ensure that their studies comply with appropriate institutional and national guidelines for ethical matters. Specifically, by submission of a manuscript, the authors are responsible for compliance with guidelines and regulations of the authors' institution and all appropriate governmental agencies.

Articles including human subjects must include a statement that informed consent was obtained and that the study was reviewed and approved by the institution's committee on human experimentation. Articles including animal experimentation must conform to the principles of the American Physiological Society, and a statement acknowledging conformation to these standards must be included

in the *Materials and methods* section of the manuscript. Authors are also requested to identify possible conflicts of interest, especially as it relates to commercial sponsorship or equity holdings.

Language

The language of the Journal is English, but acceptance of a manuscript will reflect scientific rather than grammatical content. The editors undertake to facilitate the publication of papers from those authors whose native language is not English.

Manuscripts

Manuscripts should be submitted via the web-based peer-review system, CTY Manuscript Central and must include a complete set of Figures. Further information regarding Figure formats is outlined below. Type all pages with double-spacing and wide margins on all pages, with left justified margins. Divide the manuscript into the following sections: Title Page, Abstract, Keywords (3–6), Introduction, Materials and methods, Results, Discussion, Acknowledgements, References, Tables, Figure legends and Figures.

Cover letter

A submission letter is required for all papers. A submission letter template will be attached to the confirmation email that the author will receive after completing online submission.

Authorship

Authorship should be assumed only by those workers who have contributed materially to the work and its report, and who accept the responsibility for the accuracy of the concepts expressed. Colleagues who have otherwise assisted or collaborated should be recognized in the section for acknowledgements. An excellent guide to authorship is given by the Style Manual Committee of the Council of Biology Editors, and the editors encourage consultation with this source.

Style

The Journal uses the English language, and avoids the use of Latin terms such as 'superior vena cava'.

Anatomic terms should be given in the English language. Do not use abbreviations. Headlines and subheadings should be liberally employed in the methods, results, and discussion sections. Use short paragraphs whenever possible. The authors should strive for clarity of expression, avoiding, in particular, the use of jargon. Authors should also avoid conventions such as Group 1, Group 2, and so on, using descriptive titles rather than alphanumeric codes.

Title page

Follow the on-screen instruction on CTY Manuscript Central to enter each piece. The title page should include a descriptive title, a running head title not to exceed 50 characters, authors' complete names (first names and initials followed by family name), academic addresses, corresponding author (with full address, telephone and fax numbers, and email address), keywords (3-6 for indexing purposes and words not used in main title of article), and all sources of financial support, including grants from non-profit organizations and/or any commercial support. Please note that authors' qualifications or academic positions should not be included except in the address for correspondence.

Abstract

The Abstract should be no more than 250 words and should include statements identifying relevant methods and results justifying publication. Do not use abbreviations.

Introduction

The Introduction should be brief and set out the purposes for which the study has been performed. It should not include an extensive review of the literature.

Materials and methods

The Materials and methods should be sufficiently detailed so that readers and reviewers can understand precisely what has been done without studying the references directly. The description may be abbreviated when well-accepted techniques are used with appropriate reference to previously published methods. Statements confirming conformation to institutional and governmental review of the experimental protocol (see above in the Editorial policies section) should be included here.

Results

The results should be presented precisely. Reference to Tables and Figures, to the extent that they

contribute substantively to help the reader understand clearly the relevant positive and negative findings, is encouraged.

Keep discussion of their importance to a minimum in this section of the manuscript.

Discussion

The Discussion should relate directly to the study being reported. The Discussion should interpret the results, should describe the relevance of the results, and should include a discussion of the limitations of the study. Do not include a general review of the topic.

Acknowledgements

Acknowledgements should follow the discussion.

References

References should be numbered consecutively (in superscript) as they appear in the text. Type the reference list with double-spacing on a separate sheet. References (using Index Medicus abbreviations) should appear in the style as demonstrated below. Please note that if more than six authors, the **first three** authors should be listed and then 'et al.'. Examples:

1. Redington AN, Rigby ML, Oldershaw P, Gibson DG, Shinebourne EA. Right ventricular function 10 years after the Mustard operation for transposition of the great arteries: analysis of size, shape, and wall motion. *Br Heart J* 1989; 62: 455-461.
2. Smith VR, Jones AL, Miller W et al. Left ventricular myocardial velocities in children. *Eur Heart J* 2000; 21: 104-112.
3. Zuberbuhler JR. *Clinical Diagnosis in Pediatric Cardiology*. Churchill Livingstone, New York, 1981.
4. Frantz EG. Adult respiratory distress syndrome in children. In: Harried HS, Jr (ed.) *Pediatric Pulmonary Heart Disease*. Little, Brown, Boston, 1990, pp 315-324.

Tables

Tables should follow the style as demonstrated in issues to date, and be essential to the understanding of the text. Tables should have short descriptive titles and should be numbered (1, 2, 3 etc.) as they appear sequentially in the text. If only one Table is included, it should be referred to as Table. Submit each Table on a separate sheet of paper. All abbreviations and symbols should be defined in a footnote below the Table.

Figures

Please ensure that all graphs are exclusively submitted as 2-dimensional images.

To ensure that your figures are reproduced to the highest possible standards, Cambridge Journals recommends the following formats and resolutions for supplying electronic figures.

Please ensure that your figures are saved at final publication size and are in our recommended file formats. Following these guidelines will result in high quality images being reproduced in both the print and the online versions of the journal.

Line artwork

Format: tif or eps

Colour mode: black and white (also known as 1-bit)

Resolution: 1200 dpi

Combination artwork (line/tone)

Format: tif or eps

Colour mode: grayscale (also known as 8-bit)

Resolution: 800 dpi

Black and white halftone artwork

Format: tif

Colour mode: grayscale (also known as 8-bit)

Resolution: 300 dpi

Colour halftone artwork

Format: tif

Colour mode: CMYK colour

Resolution: 300 dpi

If you require any further guidance on creating suitable electronic figures, please visit http://dx.sheridan.com/guidelines/digital_art.html. Here you will find extensive guidelines on preparing electronic figures and also have access to an online pre-flighting tool (<http://dx.sheridan.com/index.html>) where you can check if your figures are suitable for reproduction.

Case Reports / Brief Reports

Case Reports which add important new information will be published as Brief Reports. Articles published in this section should have no more than three authors, 1,000 words, an abstract (of about 60 words), 3–6 key words, two Figures or Tables, and 10 references.

Editorials and Letters to the Editor

Readers are encouraged to write about any topic that relates to cardiology in the young. Such letters will appear in *Letters to the Editor*. They should be no longer than 500 words. Editorials are written on invitation but unsolicited articles of approximately 1,500 words which may have particular topical interest will be welcomed for consideration.

Reviews

Reviews of recent developments are welcome. Authors are encouraged to contact the editor to determine the appropriateness for inclusion.

Images in Congenital Cardiac Disease

Definitive, unique, or extraordinary pictures of any aspect of congenital cardiac disease will be presented with an accompanying legend of not more than 250 words (one double-spaced page). A maximum of three authors and one reference may be included. Instructional value and artistic merit will be considered in addition to scientific import and clinical relevance.

Offprints

Article offprint order forms will be sent to the corresponding author with the proofs. If you do not receive the relevant form please email the Production Editor at: nmarshall@cambridge.org

Reprints

Article reprints (following publication) are available from rcurtis@cambridge.org. A minimum reprint order of 500 copies is required.

Permission

Requests for permission to reproduce any material originally published in *Cardiology in the Young* should be sent to the Permissions Manager at lnicol@cambridge.org

Announcements

Meetings and courses will be announced in each Issue. Refer to the Announcements Section for appropriate guidelines. Please email details to: ctyedoffice@cambridge.org. The Journals homepage, www.journals.cambridge.org/cty, offers paying advertisers the opportunity to promote conferences / courses and recruitment vacancies (subject to editorial approval). For rate details please email: ad-sales@cambridge.org

Page changes

Page changes will not be made once proofs are prepared for publication.

Articles not conforming to these instructions will be returned to the corresponding author for correction, and will delay review and publication.

(Revised April 2008)