Cardiology in the Young



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

CAMBRIDGE UNIVERSITY PRESS

```
https://doi.org/10.1017/S1047951109990588 Published online by Cambridge University Press
```

You'll discover great things here. Including yourself.

At the Cardiac Center at The Children's Hospital of Philadelphia, you'll find excellence, teamwork, renowned practitioners and some of the most challenging cases anywhere. *What better place to find out just how far you can go?*

The Cardiac Center currently has openings for: Pediatric Cardiologists in various subspecialties – board certified/board eligible Pediatric Cardiac Intensivists – board certified/board eligible Cardiac Nurse Practitioners – for both inpatient and outpatient settings Cardiac Staff Nurses – for Cardiac Intensive Care and Cardiac Care Units Echo Sonographers – with pediatric training and/or experience Physician Assistants – licensed/board certified

The Cardiac Center *at* The Children's Hospital *of* Philadelphia heart.chop.edu

For physician positions, contact Robert Shaddy, M.D., at 267-426-7518 or shaddyr@email.chop.edu. For nursing positions, contact Kim Delaney, R.N., B.S.N., at 267-426-6568 or delaney@email.chop.edu. For sonographer positions, contact Larry Barnes, at 215-590-6816 or barnesl@email.chop.edu. For physician assistant positions, contact Christy Bosler, P.A.-C., at 267-426-6021 or bosler@email.chop.edu.

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, NCHiromi Kurosawa, TokyoSupplements EditorGil Wernovsky, Philadelphia, PA

Images Editor Roxane McKay, Memphis, TN

Editorial Assistant Felicity Gil, London

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 90 1996 Forest Stewardship Council

CAMBRIDGE UNIVERSITY PRESS

Volume 19 • Number 4 • Pages 309–425

Cardiology in the Young

COPYRIGHT © 2009 CAMBRIDGE UNIVERSITY PRESS ISSN 1047-9511

Table of ContentsAugust 2009

FROM THE EDITOR-IN-CHIEF	
Short-term and long-term outcomes for congenital cardiac surgery <i>Edward Baker</i>	309
EDITORIAL	
Pulmonary hypertension associated with congenital cardiac disease Rolf M.F. Berger	311
CONTINUING MEDICAL EDUCATION	
Early postoperative care of patients with pulmonary hypertension associated with congenital cardiac disease <i>Ian Adatia, Maurice Beghetti</i>	315
ORIGINAL ARTICLES	
Functional state following the Fontan procedure Ismee A. Williams, Lynn A. Sleeper, Steven D. Colan, Minmin Lu, Elizabeth A. Stephenson, Jane W. Newburger, Welton M. Gersony, Meryl S. Cohen, James F. Cnota, Andrew M. Atz, Richard V. Williams, Renee Margossian, Andrew J. Powell, Mario P. Stylianou, Daphne T. Hsu, For the Pediatric Heart Network Investigators	320
The effect of bosentan in patients with a failing Fontan circulation Caroline Ovaert, Daisy Thijs, Daniel Dewolf, Jaap Ottenkamp, Hugues Dessy, Philip Moons, Marc Gewillig, Luc Mertens	331
Exercise capacity reflects ventricular function in patients having the Fontan circulation Katrin Klimes, Stanislav Ovroutski, Hashim Abdul-Khaliq, Peter Ewert, Vladimir Alexi-Meskishvili, Titus Kuehne, Matthias Gutberlet, Felix Berger	340
The influence over a period of 8 years of patterns of prescribing palizivumab for patients with and without congenitally malformed hearts, and in admissions to paediatric intensive care <i>Alastair Turner, Colin Begg, Benjamin Smith, Jonathan Coutts</i>	346
Common signatures for gene expression in postnatal patients with patent arterial ducts and stented arteries Peter P. Mueller, Andreas Drynda, Diane Goltz, René Hoebn, Hansjörg Hauser, Matthias Peuster	352
Analysis of regional congenital cardiac surgical outcomes in Florida using The Society of Thoracic Surgeons Congenital Heart Surgery Database Jeffrey P. Jacobs, James A. Quintessenza, Redmond P. Burke, Mark S. Bleiweis, Barry J. Byrne, Eric L. Ceithaml, William M. DeCampli, Jorge M. Giroud, Richard A. Perryman, Eliot R. Rosenkranz, Grace Wolff, Vicki Posner, Sue Steverson, William B. Blanchard, Gerry L. Schiebler	360
Is a routine chest X-ray indicated before discharge following paediatric cardiac surgery? Kerstin Bosse, Thomas Krasemann	370
Experience in a single centre with percutaneous aortic valvoplasty in children, including those with associated cardiovascular lesions David Crespo, Joaquim Miró, Suzanne J Vobecky, Nancy Poirier, Chantal Lapierre, Naicheng NZ Zhao, Nagib Dahdah	372
Retrospective study of neonatal ligation during 2002 in the United Kingdom of persistently patent arterial ducts Vidheya Venkatesh, Lleona Lee, Deborah White, Wilf Kelsall	383

Table of Contents continued

IMAGES IN CONGENITAL CARDIAC DISEASE

Rupture following aortic implantation of a right coronary artery initially arising anomalously from the pulmonary trunk	389
Daiji Takeuchi, Toshio Nakanishi, Hiromi Kurosawa	
Asymptomatic anomalous right coronary artery from the pulmonary trunk R. Thomas Collins, II, Erin Davis, Paul Stephens, Jr	391
Contrast enhanced computed tomography showing an isolated ventricular septal diverticulum <i>Farid Aliyev, Hakan Erkan, Cengizhan Türkoğlu</i>	393
Giant coronary arterial aneurysm in atypical Kawasaki disease Rebeca Sarrat Torres, Pedro Betrián Blasco, María Dolores García de la Calzada	395
Differential hyperaemia of the airways Atul Gupta, Alan Magee, Mark Rosenthal	397
Aortic aneurysms and dissection diagnosed by computed tomography 19 years after repair of coarctation <i>Arun Chandran, Mark S. Bleiweis, F. Jay Fricker</i>	398
BRIEF REPORTS	
Percutaneous closure of a patent arterial duct in a newborn Fredy H. Prada, Juan M. Carretero, Lorenzo Jimenez	400
Surgical repair in neonatal life of cardiac haemangiomas diagnosed prenatally Jayendra Sharma, Yasutaka Hirata, Ralph S. Mosca	403
Successful use of intravenous amiodarone in a child with combined postoperative junctional and ectopic tachycardias	407
Mehmet Karacan, Haşim Olgun, Necip Becit	
Pulmonary diffuse arterial calcifications: a very rare complication in the recipient of a twin-to-twin transfusion syndrome <i>Rania Bassil Eter, Yves Dulac, Philippe Acar</i>	410
Incessant pericardial effusion in a 9 year old male responding to infliximab Rowan F. Walsh, Devyani Chowdhury	413
LETTERS TO THE EDITOR	
The role of echocardiography in diagnosing carditis in the setting of acute rheumatic fever John Lawrenson, Liesl Zühlke, Rik De Decker	416
The role of echocardiography in diagnosing carditis in the setting of acute rheumatic fever <i>Ishwarappa B. Vijayalakshmi</i>	417
The role of echocardiography in diagnosing carditis in the setting of acute rheumatic fever Alessandra Benettoni, Emanuela Berton, Angela De Cunto, Andrea Taddio, Loredana Lepore	419
The role of echocardiography in diagnosing carditis in the setting of acute rheumatic fever <i>Ishwarappa B. Vijayalakshmi</i>	421
NEWS FROM THE ASSOCIATION FOR EUROPEAN PAEDIATRIC CARDIOLOGY	422

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 320107; Fax: +44 (0)1223 3215070; 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 £209 or US\$475; Institutional rate £565 or US\$493.0AII 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 letture, review, or thesis; (b) that it is not under consideration for publication elsewhere; (c) that all co-authors approve its publication; (d) that is 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 garee to the automatic transfer of the copyright to the opyright 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 are biographically or stored on microfilm, in video disks, etc., without first obtaining written permission from the publisher; for the advertiser to comply with all elag requirements, 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

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 webbased 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:

- 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.
- Zuberbuhler JR. Clinical Diagnosis in Pediatric Cardiology. Churchill Livingstone, New York, 1981.
- 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 preflighting 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)