

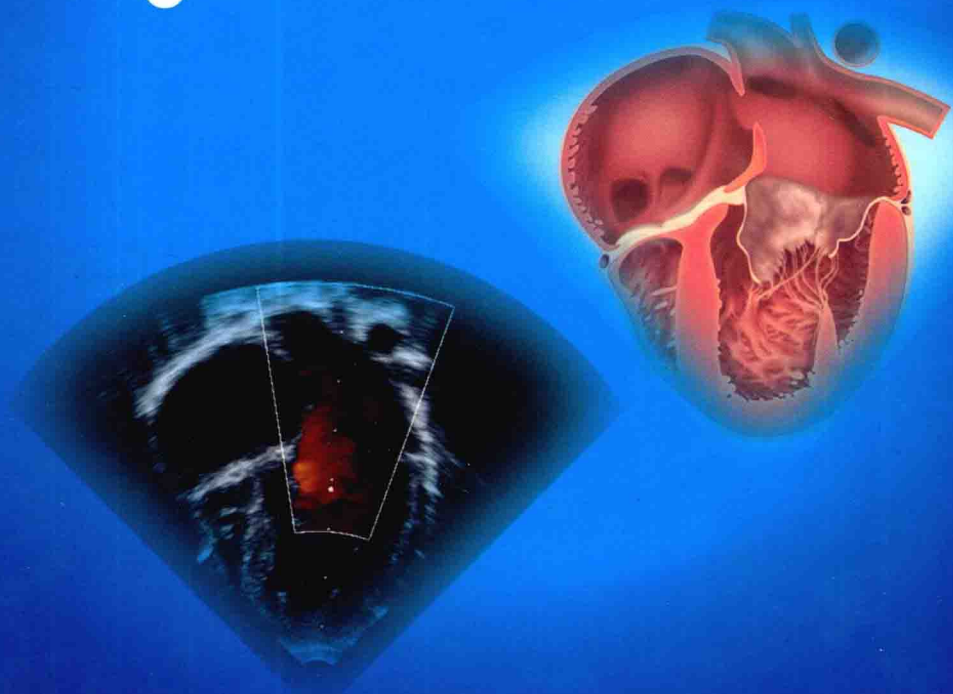
Expert **CONSULT**

Activate at [expertconsult.com](http://expertconsult.com)

Searchable Full  
Text Online

**LEWIN • STOUT**

# Echocardiography in Congenital Heart Disease



**PRACTICAL ECHOCARDIOGRAPHY SERIES**  
SERIES EDITOR: CATHERINE M. OTTO

**ELSEVIER**  
SAUNDERS

# Echocardiography in Congenital Heart Disease

PRACTICAL ECHOCARDIOGRAPHY SERIES

No part of this publication may be reproduced or transmitted in any form or by any means, electronic or mechanical, including photocopying, recording, or any information storage and retrieval system, without permission in writing from the publisher. Details on how to seek permission, further information about the Publisher's permissions policies and our arrangements with organizations such as the Copyright Clearance Center and the Copyright Licensing Agency, can be found at our website: [www.elsevier.com/permissions](http://www.elsevier.com/permissions).

This book and the individual contributions contained in it are protected under copyright by the Publisher (other than as may be noted herein).

#### Notices

Knowledge and best practice in this field are constantly changing. As new research and experience broaden our understanding, changes in research methods, professional practices, or medical treatment may become necessary.

Practitioners and researchers must always rely on their own experience and knowledge in evaluating and using any information, methods, compounds, or experiments described herein. In using such information or methods they should be mindful of their own safety and the safety of others, including parties for whom they have a professional responsibility.

With respect to any drug or pharmaceutical products identified, readers are advised to check the most current information provided (i) on procedures featured or (ii) by the manufacturer of each product to be administered, to verify the recommended dose or formula, the method and duration of administration, and contraindications. It is the responsibility of practitioners, relying on their own experience and knowledge of their patients, to make diagnoses, to determine dosages and the best treatment for each individual patient, and to take all appropriate safety precautions.

To the fullest extent of the law, neither the Publisher nor the authors, contributors, or editors, assume any liability for any injury and/or damage to persons or property as a matter of products liability, negligence or otherwise, or from any use or operation of any methods, products, instructions, or ideas contained in the material herein.

#### Library of Congress Cataloging-in-Publication Data

Echocardiography in congenital heart disease/[edited by] Mark B. Lewin, Karen Stout.

p. ; cm.—(Practical echocardiography series)

Includes bibliographical references and index.

ISBN 978-1-4377-2696-1 (hardcover : alk. paper)

I. Lewin, Mark B. II. Stout, Karen. III. Series: Practical echocardiography series.

[DNLM: 1. Echocardiography—methods—Handbooks. 2. Heart Defects, Congenital—ultrasonography—Handbooks. WG 39]

LC classification not assigned

618.92'1207543—dc23

2011036498

*Senior Acquisitions Editor:* Dolores Meloni  
*Editorial Assistant:* Brad McIlwain  
*Publishing Services Manager:* Pat Joiner-Myers  
*Project Manager:* Marlene Weeks  
*Designer:* Steven Stave

Printed in Canada.

Last digit is the print number: 9 8 7 6 5 4 3 2 1

Working together to grow  
libraries in developing countries

[www.elsevier.com](http://www.elsevier.com) | [www.bookaid.org](http://www.bookaid.org) | [www.sabre.org](http://www.sabre.org)

ELSEVIER

BOOK AID  
International

Sabre Foundation

# **Look for these other titles in Catherine M. Otto's Practical Echocardiography Series**

---

**Donald C. Oxorn**

*Intraoperative Echocardiography*

**Linda D. Gillam & Catherine M. Otto**

*Advanced Approaches in Echocardiography*

**Martin St. John Sutton & Susan E. Wiegers**

*Echocardiography in Heart Failure*



# Echocardiography in Congenital Heart Disease

PRACTICAL ECHOCARDIOGRAPHY SERIES

**Mark B. Lewin, MD**

Professor and Chief  
Division of Pediatric Cardiology  
University of Washington School of Medicine  
Heart Center Co-Director and Director of Pediatric Echocardiography  
Seattle Children's Hospital  
Seattle, Washington

**Karen Stout, MD**

Director, Adult Congenital Heart Disease Program  
Associate Professor, Departments of Medicine and Pediatrics  
University of Washington School of Medicine  
Attending Cardiologist  
University of Washington Medical Center and Seattle Children's Hospital  
Seattle, Washington

ELSEVIER  
SAUNDERS

# Contributors

## **Peter J. Cawley, MD, FACC**

Acting Assistant Professor of Medicine, University of Washington School of Medicine; Attending Cardiologist, University of Washington Medical Center, Seattle, Washington  
*Thromboembolic Phenomena and Vegetations*

## **Nadine F. Choueiter, MD**

Pediatric Cardiology Fellow, University of Washington School of Medicine; Seattle Children's Hospital, Seattle, Washington  
*Echocardiographic Imaging of Single-Ventricle Lesions*

## **Raylene M. Choy, RDCS**

Cardiac Sonographer, Heart Center, Seattle Children's Hospital, Seattle, Washington  
*Echocardiographic Imaging of Single-Ventricle Lesions*

## **Jeffrey A. Conwell, MD**

Associate Professor of Pediatrics, Division of Pediatric Cardiology, University of Washington School of Medicine; Attending Cardiologist, Seattle Children's Hospital, Seattle, Washington  
*Atrioventricular Septal Defect: Echocardiographic Assessment*  
*Kawasaki Disease: Echocardiographic Assessment*

## **Brandy Hattendorf, MD**

Assistant Professor of Pediatrics, University of Washington School of Medicine; Attending Cardiologist, Seattle Children's Hospital, Seattle, Washington  
*Shunting Lesions*  
*Implications of Pediatric Renal, Endocrine, and Oncologic Disease*

## **Denise Joffe, MD**

Associate Professor of Anesthesiology, Department of Anesthesiology, University of Washington School of Medicine; Seattle Children's Hospital, Seattle, Washington  
*Intraoperative Transesophageal Echocardiography*

## **Troy Johnston, MD**

Associate Professor of Pediatrics, University of Washington School of Medicine; Attending Cardiologist, Seattle Children's Hospital, Seattle, Washington  
*Echocardiography in the Cardiac Catheterization Laboratory*

## **Mariska Kemna, MD**

Assistant Professor of Pediatrics, University of Washington School of Medicine; Attending Cardiologist, Seattle Children's Hospital, Seattle, Washington  
*Myocardial Pathology*  
*Echocardiographic Assessment After Heart Transplantation*

## **Joel Lester, RDCS**

Echocardiography Laboratory Supervisor, Heart Center, Seattle Children's Hospital, Seattle, Washington  
*The Pediatric Transthoracic Echocardiogram*

## **Mark B. Lewin, MD**

Professor and Chief, Division of Pediatric Cardiology, University of Washington School of Medicine; Heart Center Co-Director and Director of Pediatric Echocardiography, Seattle Children's Hospital, Seattle, Washington  
*The Pediatric Transthoracic Echocardiogram*  
*Right Heart Anomalies*

**Maggie L. Likes, MD**

Assistant Professor of Pediatrics, University of Washington School of Medicine; Attending Cardiologist, Seattle Children's Hospital, Seattle, Washington  
*Right Heart Anomalies*

**David S. Owens, MD**

Acting Assistant Professor of Medicine, University of Washington School of Medicine; Attending Cardiologist, University of Washington Medical Center, Seattle, Washington  
*Left Heart Anomalies*  
*Myocardial Pathology*

**Amy H. Schultz, MD**

Assistant Professor of Pediatrics, University of Washington School of Medicine; Attending Cardiologist, Seattle Children's Hospital, Seattle, Washington  
*Conotruncal Lesions*  
*Transposition of the Great Arteries*

**Brian D. Soriano, MD**

Assistant Professor of Pediatrics, University of Washington School of Medicine; Attending Cardiologist, Seattle Children's Hospital, Seattle, Washington  
*Venous Anomalies*  
*Left Heart Anomalies*  
*Thromboembolic Phenomena and Vegetations*

**Karen Stout, MD**

Director, Adult Congenital Heart Disease Program, Associate Professor, Departments of Medicine and Pediatrics, University of Washington School of Medicine; Attending Cardiologist, University of Washington Medical Center and Seattle Children's Hospital, Seattle, Washington

**Margaret M. Vernon, MD**

Assistant Professor of Pediatrics, University of Washington School of Medicine; Attending Cardiologist, Seattle Children's Hospital, Seattle, Washington  
*The Fetal Echocardiogram*



# Foreword

Echocardiography is a core component of every aspect of clinical cardiology and now plays an essential role in daily decision making. Both echocardiographers and clinicians face unique challenges in interpretation of imaging and Doppler data and in integration of these data with other clinical information. However, with the absorption of echocardiography into daily patient care, there are several unmet needs in our collective knowledge base. First, clinicians caring for patients need to understand the value, strengths, and limitations of echocardiography relevant to their specific scope of practice. Second, echocardiographers need a more in-depth understanding of the clinical context of the imaging study. Finally, there often are unique aspects of data acquisition and analysis in different clinical situations, all of which are essential for accurate echocardiographic diagnosis. The books in the *Practical Echocardiography Series* are aimed at filling these knowledge gaps, with each book focusing on a specific clinical situation in which echocardiographic data are key for optimal patient care.

In addition to *Echocardiography in Congenital Heart Disease*, edited by Mark B. Lewin, MD, and Karen Stout, MD, other books in the series are *Intraoperative Echocardiography*, edited by Donald C. Oxorn, MD; *Echocardiography in Heart Failure*, edited by Martin St. John Sutton, MD, and Susan E. Wieggers, MD; and *Advanced Approaches in Echocardiography*, edited by Linda D. Gillam, MD, and myself. Information is presented as concise bulleted text accompanied by numerous illustrations and tables, providing a practical approach to data acquisition and analysis, including technical details, pitfalls, and clinical interpretation, supplemented by web-based video case examples. Each volume in this series expands on the basic

principles presented in the *Textbook of Clinical Echocardiography*, fourth edition, and can be used as a supplement to that text or can be used by physicians interested in a focused introduction to echocardiography in their area of clinical practice.

Patients with congenital heart disease are increasingly encountered in clinical practice due to the success of surgical and medical treatment of these conditions, allowing survival into adulthood. At the same time, each of us may see only a few cases of each type of congenital heart disease because of the wide range of congenital lesions and the variety of surgical repair techniques. Thus, easily accessible and concise information is needed when these patients are seen to ensure that the echocardiographic study is performed and interpreted correctly.

The editors of *Echocardiography in Congenital Heart Disease*, Mark B. Lewin and Karen Stout, are recognized experts with robust clinical experience that includes pediatric, adolescent, and adult patients with congenital heart disease. In this book the editors provide a comprehensive discussion of echocardiography in the patient with congenital heart disease, spanning the entire age range from birth to old age. This book is aimed at all clinicians who care for patients with congenital heart disease, whether in the pediatric or adult setting, including cardiologists, cardiology fellows, cardiac sonographers, anesthesiologists, and cardiac surgeons.

The wealth of information provided in this book is truly awesome. Every clinician who sees patients with congenital heart disease and every echocardiography laboratory will want a copy close at hand.

Catherine M. Otto, MD



# Preface

This text is one of four in the *Practical Echocardiography Series*, which covers the range of echocardiographic topics. The topics in the other three volumes in this set include *Intraoperative Echocardiography*, *Echocardiography in Heart Failure*, and *Advanced Approaches in Echocardiography*. This volume provides a resource for those interested in pediatric and adult congenital echocardiography. The chapters are designed to review basic principles, provide details of image acquisition and interpretation, and describe how echocardiography is used to develop management strategies.

This book will be of interest to cardiology and sonographer trainees, as well as practicing cardiologists and sonographers, as an overview of pediatric and congenital echocardiography. The chapters cover general pediatric echo imaging protocols, individual congenital cardiac diagnoses, cardiomyopathies, and other pediatric organ system disorders in which cardiac structural or functional assessment is necessary. There are also

chapters devoted to congenital transesophageal echo as well as echo imaging in the cardiac catheterization laboratory.

Each chapter includes a *step-by-step approach to patient examination*, bulleted points of *major principles*, and lists of *key points*. Those areas where echo can serve as a resource for accurately working through a differential diagnosis are also pointed out. Methods regarding quantitative data analysis and calculations are also included. Numerous echo images and illustrations with detailed figure legends demonstrate important principles. This book does not replace formal training in pediatric and congenital echocardiography but rather serves as a supplement to this training. Accredited training is the only method of obtaining all the tools needed to obtain accurate echocardiographic data, and we fully endorse this process.

**Mark B. Lewin, MD**  
**Karen Stout, MD**

# Acknowledgments

We could never have completed this work if not for the dedication and skills of our authors. The cardiac sonographers at Seattle Children's Hospital and the University of Washington deserve recognition for their commitment to superb imaging and the dedication they show to patients, families, and their colleagues. From Seattle Children's these include Heidi Borchers, RDCS; Colleen Cailles, RDCS; Raylene Choy, RDCS; Mikki Clouse, RDCS; Judy Devine, RDCS; Alison Freeberg, RDCS; Laura Huntley, RDCS; Mary Jordan, RDCS; Joel Lester, RDCS; Danielle Saliba, RDCS; Pauline Suon, RDCS; Shelby Thomas-Irish, RDCS; and Erin Trent, RDCS. From the University of Washington these include Caryn D'Jang, RDCS; Michelle Fujioka, RDCS; Yelena Kovalenko, RDCS; Amy Loscher, RDCS; Todd

Zwink, RDCS; Pamela Clark, RDCS; Sarah Curtis, RDCS; Jennifer Gregov, RDCS; Carol Kraft, RDCS; Chris McKenzie, RDCS; Joannalyn Sangco, RDCS; and Rebecca G. Schwaegler, RDCS. Special thanks to Catherine Otto, MD, for her careful attention to detail and dedication to this project. We also wish to acknowledge Natasha Andjelkovic, Bradley McIlwain, and Marla Sussman at Elsevier, who kept us on track and on time.

Of course, finally (and most importantly) the unwavering support of our families cannot be overlooked. Deb, Johanne, Julien, and Cal are always in our hearts!

**Mark B. Lewin, MD**  
**Karen Stout, MD**

# Glossary

- 2C two-chamber view  
4C four-chamber view  
5C five-chamber view  
2D two-dimensional  
3D three-dimensional  
A4C apical four-chamber view  
AA aortic arch  
AAO aortic arch obstruction  
ACC American College of Cardiology  
AHA American Heart Association  
AI aortic insufficiency  
ALCAPA anomalous origin of the left coronary artery from the pulmonary artery  
Ao aorta  
APB absent pulmonary valve  
AR aortic regurgitation  
aRV atrialized right ventricle  
AS aortic stenosis; aortic septum  
ASD atrial septal defect  
ASO arterial switch operation  
AV atrioventricular; aortic valve  
AVC atrioventricular canal; aortic valve closure  
AVR aortic valve replacement  
AVS atrioventricular septum  
AVSD atrioventricular septal defect  
AVV atrioventricular valve  
AVVR atrioventricular valve regurgitation  
bpm beats per minute  
BSA body surface area  
BT Blalock-Taussig  
BVF bulboventricular foramen  
cc-TGA congenitally corrected transposition of the great arteries  
CFD color flow Doppler  
CHD congenital heart disease  
CI confidence interval  
CM cardiomyopathy  
CMR cardiac magnetic resonance imaging  
CoA coarctation of the aorta  
CPB cardiopulmonary bypass  
CS coronary sinus  
CT computed tomography  
CW continuous wave  
Cx circumflex coronary artery  
DA ductus arteriosus  
DAo descending aorta  
DCM dilated cardiomyopathy  
DCRV double-chamber right ventricle  
DILV double-inlet left ventricle  
DKS Damus-Kaye-Stansel (procedure)  
DORV double-outlet right ventricle  
 $dp/dt$  rate of change in pressure over time  
DSE dobutamine stress echocardiography  
 $dT/dt$  rate of increase in temperature  
d-TGA dextro-transposition of the great arteries  
E early diastolic peak velocity  
E' early diastolic tissue Doppler velocity  
ECG electrocardiogram  
echo echocardiography  
EF ejection fraction  
EFE endocardial fibroelastosis  
ET ejection time  
FAC fractional area change  
FO foramen ovale  
FS fractional shortening  
GOS Great Ormond Street  
GV great vessel  
HCM hypertrophic cardiomyopathy  
HIV human immunodeficiency virus  
HLHS hypoplastic left heart syndrome  
HR heart rate  
IAS interatrial septum  
ICE intracardiac echocardiography  
ILB inferior limbic bands  
IVA isovolumic acceleration  
IVC inferior vena cava; isovolumic contraction  
IVCT isovolumic contraction time  
IVRT isovolumic relaxation time  
IVS interventricular septum; intact ventricular system  
IVSD inlet ventricular septal defect  
LA left atrium  
LAA left atrial appendage  
LAD left descending artery  
LAE left atrial enlargement  
LAX long axis view  
LCA left coronary artery  
LCC left coronary cusp



- LLPV** left lower pulmonary vein  
**LM** left mural leaflet  
**LMCA** left main coronary artery  
**LPA** left pulmonary artery  
**LRV** lower reference value  
**LSVC** left superior vena cava  
**L-TGA** levo-TGA  
**LUPV** left upper pulmonary vein  
**LV** left ventricle  
**LVE** left ventricular enlargement  
**LVED** left ventricular end-diastolic dimension  
**LVH** left ventricular hypertrophy  
**LVM** left ventricular mass  
**LVN** left ventricular noncompaction  
**LVO** left ventricular outflow  
**LVOT** left ventricular outflow tract  
**LVOTO** left ventricular outflow tract obstruction  
**LVP** left ventricular pressure  
**LVPW** left ventricular posterior wall  
**MAPCA** major aortopulmonary collateral artery  
**m-BT** modified Blalock-Taussig  
**ME** midesophageal  
**MIPG** maximal instantaneous pressure gradient  
**M-mode** motion display (depth versus time)  
**MPA** main pulmonary artery  
**MPI** myocardial performance index  
**MPR** multiplanar reconstruction  
**MR** mitral regurgitation  
**MRI** magnetic resonance imaging  
**MS** mitral stenosis  
**MV** mitral valve  
**MVI** myocardial videointensity  
**NBTE** nonbacterial thrombotic endocarditis  
**NCC** noncoronary cusp  
**PA** pulmonary artery; pulmonary atresia  
**PA/IVS** pulmonary atresia with intact ventricular septum  
**PAP** pulmonary artery pressure  
**PAPVC** partial anomalous pulmonary venous connection  
**PAPVD** partial anomalous pulmonary venous drainage  
**PASP** pulmonary artery systolic pressure  
**PBF** pulmonary blood flow  
**PBL** posterior bridging leaflet  
**PDA** patent ductus arteriosus; posterior descending artery  
**PE** pericardial effusion  
**PFO** patent foramen ovale  
**PHTN** pulmonary hypertension  
**PI** pulmonary insufficiency  
**PIPG** peak instantaneous pressure gradient  
**PISA** proximal isovelocity surface area  
**PLAX** parasternal long axis view  
**PPV** positive pressure ventilation  
**PR** pulmonary regurgitation  
**PS** pulmonary stenosis  
**PSAX** parasternal short axis view  
**pulmV** pulmonary valve  
**PV** pulmonary vein  
**PVR** pulmonary vascular resistance  
**PW** pulsed wave  
 $Q_p$  pulmonary volume flow rate  
 $Q_s$  systemic volume flow rate  
**RA** right atrium  
**RAA** right atrial appendage  
**RAE** right atrial enlargement  
**RAP** right atrial pressure  
**RCA** right coronary artery  
**RCC** right coronary cusp  
**RCM** restrictive cardiomyopathy  
**RI** right inferior leaflet  
**RLPV** right lower pulmonary vein  
**ROA** regurgitant orifice area  
**RPA** right pulmonary artery  
**RUPV** right upper pulmonary vein  
**RV** right ventricle  
**RVD** right ventricle diameter  
**RVDCC** right ventricle-dependent coronary circulation  
**RVE** right ventricle enlargement  
**RVEDV** right ventricular end-diastolic volume  
**RVH** right ventricular hypertrophy  
**RVO** right ventricular outflow  
**RVOT** right ventricular outflow tract  
**RVOTO** right ventricular outflow tract obstruction  
**RVP** right ventricular pressure  
**RWMA** regional wall motion abnormality  
**s** second  
**SAM** systolic anterior motion  
**SAX** short axis view  
**SBF** systemic blood flow  
**SC** subcostal  
**SCD** sudden cardiac death  
**SCLAX** subcostal long axis view  
**SCSAX** subcostal short axis view  
**SD** standard deviation  
**SLB** superior limbic band  
**SLE** systemic lupus erythematosus  
**SR** strain rate  
**SSFP** single-state free-precession  
**SSN** suprasternal notch  
**SV** single ventricle; stroke volume  
**SVC** superior vena cava  
**TA** tricuspid atresia  
**TAPSE** tricuspid annular plane systolic excursion  
**TAPVC** total anomalous pulmonary venous connection  
**TAPVD** total anomalous pulmonary venous drainage  
**TDI** tissue Doppler imaging  
**TEE** transesophageal echocardiography  
**TG** transgastric  
**TGA** transposition of the great arteries



**TOF** tetralogy of Fallot  
**TR** tricuspid regurgitation  
**TS** tricuspid stenosis  
**TTE** transthoracic echocardiography  
**TV** tricuspid valve

**UE** upper esophageal  
**URV** upper reference value  
**VS** ventricular septum  
**VSD** ventricular septal defect  
**VVI** velocity vector imaging

# Contents

## **I** PEDIATRIC AND CONGENITAL IMAGING PRINCIPLES AND TECHNIQUES

- 1** The Pediatric Transthoracic Echocardiogram 3  
*Joel Lester and Mark B. Lewin*

- 2** The Fetal Echocardiogram 14  
*Margaret M. Vernon*

- 3** Echocardiography in the Cardiac Catheterization Laboratory 22  
*Troy Johnston*

- 4** Intraoperative Transesophageal Echocardiography 31  
*Denise Joffe*

## **II** CONGENITAL HEART DISEASE

- 5** Shunting Lesions 61  
*Brandy Hattendorf*

- 6** Atrioventricular Septal Defect: Echocardiographic Assessment 74  
*Jeffrey A. Conwell*

- 7** Conotruncal Lesions 84  
*Amy H. Schultz*

- 8** Transposition of the Great Arteries 104  
*Amy H. Schultz*

- 9** Venous Anomalies 124  
*Brian D. Soriano*

- 10** Echocardiographic Imaging of Single-Ventricle Lesions 132  
*Nadine F. Choueiter and Raylene M. Choy*

- 11** Right Heart Anomalies 145  
*Maggie L. Likes and Mark B. Lewin*

- 12** Left Heart Anomalies 169  
*Brian D. Soriano and David S. Owens*

- 13** Myocardial Pathology 183  
*David S. Owens and Mariska Kemna*

## **III** ACQUIRED HEART DISEASE IN THE CHILD

- 14** Kawasaki Disease: Echocardiographic Assessment 199  
*Jeffrey A. Conwell*

**15 Thromboembolic Phenomena  
and Vegetations 206**

*Peter J. Cawley and Brian D. Soriano*

**16 Implications of Pediatric Renal,  
Endocrine, and Oncologic  
Disease 215**

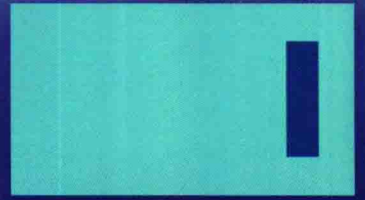
*Brandy Hattendorf*

**17 Echocardiographic Assessment  
After Heart**

**Transplantation 224**

*Mariska Kemna*

**Index 235**



# **PEDIATRIC AND CONGENITAL IMAGING PRINCIPLES AND TECHNIQUES**



