

PEDIATRIC CARDIAC SURGERY

FIFTH EDITION

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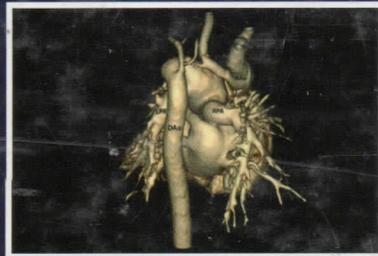
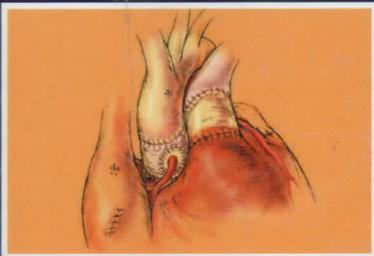
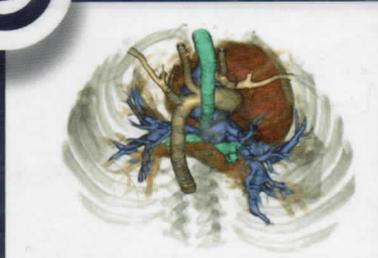
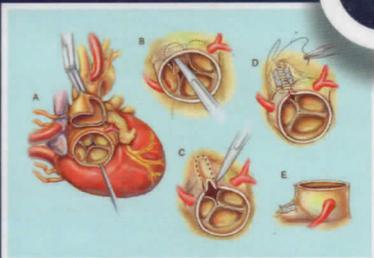
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Pediatric Cardiac Surgery

FIFTH EDITION

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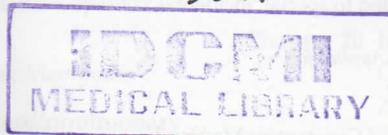
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3365



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M
617. 412059
P341

This fifth edition first published 2023
© 2023 by John Wiley & Sons Ltd.

Edition History
John Wiley & Sons (4e, 2013)
Mosby (3e, @2003; 2e, 1994)

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Library of Congress Cataloging-in-Publication Data is Applied for
Hardback ISBN: 9781119282310

Cover image: © Pobytov/Getty Images; Courtesy of Constantine Mavroudis
Cover design by Wiley

Set in 9.25/12pt Palatino by Straive, Chennai, India

Printed in Singapore

M067483_110123

Table of Contents

Contributors, v

Preface to the Fifth Edition, x

A Note on Nomenclature, xii

Chapter 1 Development of the Heart and Great Vessels, 1
Ram Kumar Subramanyan, Robert H. Anderson, and Peter J. Gruber

Chapter 2 Genetics of Congenital Heart Disease, 25
Peter J. Gruber

Chapter 3 Fetal Cardiac Physiology and Fetal Cardiac Intervention, 35
Timothy S. Lancaster, Jacob R. Miller, and Pirooz Eghtesady

Chapter 4 Preoperative Diagnostic Evaluation, 47
Barbara J. Deal, Amanda L. Hauck, Sabrina Tsao, Angira Patel, Simon Lee, and Doff B. McElhinney

Chapter 5 Hybrid Procedures for Congenital Heart Disease, 87
Hakan Akintuerk, Dietmar Schranz, and Norbert Voelkel

Chapter 6 Anesthesia for the Patient with Congenital Heart Disease, 99
H. Jay Przybylo

Chapter 7 Perioperative Care, 113
Mjaye L. Mazwi, Carl L. Backer, John M. Costello, and Constantine Mavroudis

Chapter 8 Palliative Operations, 143
Carl L. Backer and Constantine Mavroudis

Chapter 9 Management of Pediatric Cardiopulmonary Bypass, 161
Nicholas D. Andersen, James M. Meza, and Joseph W. Turek

Chapter 10 Pediatric Myocardial Protection, 191
Sachin Talwar and Shiv Kumar Choudhary

Chapter 11 Patent Arterial Duct, 213
Elizabeth H. Stephens, Paul Tannous, Carl L. Backer, and Constantine Mavroudis

Chapter 12 Vascular Rings and Pulmonary Artery Sling, 225
Carl L. Backer, Cynthia K. Rigsby, and Constantine Mavroudis

Chapter 13 Coarctation of the Aorta, 249
Carl L. Backer, Joseph A. Dearani, and Constantine Mavroudis

Chapter 14 Interrupted Aortic Arch, 279
Dilip S. Nath and Richard A. Jonas

Chapter 15 Atrial Septal Defect, Partial Anomalous Pulmonary Venous Connection, and Scimitar Syndrome, 299
Carl L. Backer, Paul Tannous, and Constantine Mavroudis

Chapter 16 Ventricular Septal Defect, 317
Constantine Mavroudis, Carl L. Backer, and Robert H. Anderson

Chapter 17 Atrioventricular Septal Defects, 361
Carl L. Backer and Constantine Mavroudis

Chapter 18 Common Arterial Trunk, 383
Constantine Mavroudis and Carl L. Backer

Chapter 19 Aortopulmonary Window and Aortic Origin of a Pulmonary Artery, 409
Stephanie Fuller and Robert H. Anderson

Chapter 20 Isolated Right Ventricular Outflow Tract Obstruction, 419
Ali Dodge-Khatami and Christopher E. Greenleaf

Chapter 21 Tetralogy of Fallot, 431
Ali Dodge-Khatami, Peter Chen, and Constantine Mavroudis

- Chapter 22** Tetralogy of Fallot with Pulmonary Atresia and Major Aortopulmonary Collaterals, 463
Matthew Liava'a and Yves d'Udekem
- Chapter 23** Ventricular to Pulmonary Artery Conduits, 481
John W. Brown and Jeremy L. Herrmann
- Chapter 24** Double-Outlet Right Ventricle, 499
Constantine Mavroudis, Carl L. Backer, and Robert H. Anderson
- Chapter 25** Transposition of the Great Arteries, 539
Constantine Mavroudis, Carl L. Backer, and Jeremy L. Herrmann
- Chapter 26** Congenitally Corrected Transposition of the Great Arteries, 581
Tom R. Karl and Jeffrey P. Jacobs
- Chapter 27** The Functionally Univentricular Heart, 601
Peter Sassalos, Ming-Sing Si, Jennifer C. Romano, Edward L. Bove, and Richard G. Ohye
- Chapter 28** Fontan Conversion, 629
Constantine Mavroudis, Barbara J. Deal, and Carl L. Backer
- Chapter 29** Ebstein Malformation, 649
Kimberly A. Holst and Joseph A. Dearani
- Chapter 30** Left Ventricular Outflow Tract Obstruction, 669
William M. DeCampli and Kamal K. Pourmoghadam
- Chapter 31** Hypertrophic Cardiomyopathy, 705
William M. DeCampli and Kamal K. Pourmoghadam
- Chapter 32** Hypoplastic Left Heart Syndrome, 719
Chun Soo Park and James S. Tweddell
- Chapter 33** Aorto-Left Ventricular Tunnel, 743
Jeremy L. Herrmann and Stephanie Fuller
- Chapter 34** Congenital Anomalies of the Mitral Valve, 749
Perry S. Choi and Sitaram M. Emani
- Chapter 35** Total Anomalous Pulmonary Venous Connection, 771
Rachel D. Vanderlaan and Christopher A. Caldarone
- Chapter 36** *Cor Triatriatum*, Pulmonary Vein Stenosis, and Atresia of the Common Pulmonary Vein, 787
Constantine D. Mavroudis, Robert H. Anderson, and Constantine Mavroudis
- Chapter 37** Anomalous Systemic Venous Connections, 801
Henry L. Walters III and Ralph E. Delius
- Chapter 38** Connective Tissue Disorders, 821
Charles D. Fraser III, Trevor A. Ellison, Duke E. Cameron, and Luca A. Vricella
- Chapter 39** Coronary Artery Anomalies, 835
Constantine Mavroudis, Ali Dodge-Khatami, and Carl L. Backer
- Chapter 40** Cardiac Tumors, 867
Rüdiger Lange and Thomas Günther
- Chapter 41** Diseases of the Pericardium, 883
Elizabeth H. Stephens and Victor O. Morell
- Chapter 42** Surgical and Transcatheter Management of Arrhythmias, 895
Barbara J. Deal and Constantine Mavroudis
- Chapter 43** Pediatric Heart Transplantation, 921
Charles B. Huddleston and Andrew C. Fiore
- Chapter 44** Lung and Heart-Lung Transplantation, 941
Charles B. Huddleston and Andrew C. Fiore
- Chapter 45** Endocarditis in Patients with Congenital Heart Disease, 957
Peter D. Wearden and Constantine Mavroudis
- Chapter 46** Pediatric Mechanical Circulatory Support, 983
Michelle Ploutz, Angela Lorts, and David L.S. Morales
- Chapter 47** Adult Congenital Heart Disease, 999
Maria Drakopoulou, Konstantinos Dimopoulos, Stamatia Prapa, Darryl F. Shore, Stella Brili, and Michael Gatzoulis
- Chapter 48** Bioethics in Congenital Heart Surgery, 1055
Constantine Mavroudis, Constantine D. Mavroudis, Thomas Cook, Catherine L. Mavroudis, Allison Siegel, and Alex Golden
- Chapter 49** Education in Congenital Cardiac Surgery, 1087
Constantine D. Mavroudis, Constantine Mavroudis, Carl L. Backer, and Richard H. Feins
- Index, 1095

Development of the Heart and Great Vessels

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Major advances in molecular genetics, establishment of appropriate animal models, and improvements in analytical techniques have contributed to a greater understanding of cardiac development. Modern cardiac embryology now combines molecular and cellular biologic techniques with traditional embryologic morphologic approaches across multiple model systems. A significant proportion of our understanding continues to be derived from nonhuman experimental models, supplemented by observations imputed from the congenitally malformed human heart [1]. In early studies, avian embryos were the favored experimental model because of the ease with which they could be observed and manipulated. Due to the strength of genetic and molecular investigative tools, the mouse has now become the preferred model for studying cardiac development. Table 1.1 provides a simplified comparison of developmental staging in human, mouse, and chicken embryos [2–8]. Understanding cardiac development not only has implications for classifying and managing congenital heart disease, but also provides a platform for the development of novel management approaches, in both children and adults.

With a goal of simplifying the description of complex developmental structures, in this chapter we have made efforts to harmonize nomenclature using descriptive terms that relate as much as possible to human development. Thus, "anterior-posterior axis" is replaced by "dorsal-ventral axis" or "cranial-caudal." "Anterior" is often replaced by "ventral" or "cranial." "Posterior" is frequently replaced by "dorsal" or "caudal." "Conus" is replaced by "proximal outflow tract," and "truncus" is replaced by "distal outflow tract." The "dorsal mesocardial protrusion" is referred to as the "vestibular spine."

Origin of Cardiac Precursor Cells

All cells destined to become part of the heart derive from populations of undifferentiated precursors. These precursor cells are influenced by external signals and guided to their final developmental state. In humans, during the second week following fertilization, the blastocyst has partially embedded into the uterine endometrium. At this stage, the inner cell mass, or embryoblast, differentiates into two distinct layers of cells: a larger columnar epiblast layer and the smaller cuboidal hypoblast layer. The third week of development is characterized by the next critical embryonic process, termed gastrulation. A primitive streak is formed in the epiblast layer, following which some epiblast cells invaginate under and displace the hypoblast. Subsequent widespread cell migration into, and reorganization within, the blastocoel cavity results in the formation of three germ layers: the ectoderm, mesoderm, and endoderm. This sets the stage for the determination of the future body plan of the embryo (Figure 1.1) [3,9].

Migrating epiblast cells, which have now formed the mesoderm of the embryo, gradually travel cephalad. During this migration, they join the lateral plate mesoderm at the level of the primitive node. The lateral plate mesoderm then divides into two layers: a splanchnic layer directly above the endoderm, and a somatic layer directly below the ectoderm. The anterior endoderm provides signals to splanchnic mesodermal cells to enter the precardiac lineage. Fibroblast growth factors (FGFs)-1, -2, and -4, and bone morphogenetic protein 2 (BMP-2), are proteins that appear to be critical to this process [10]. To date, however, no single gene has been identified whose ablation leads to a specific failure of all myocardial