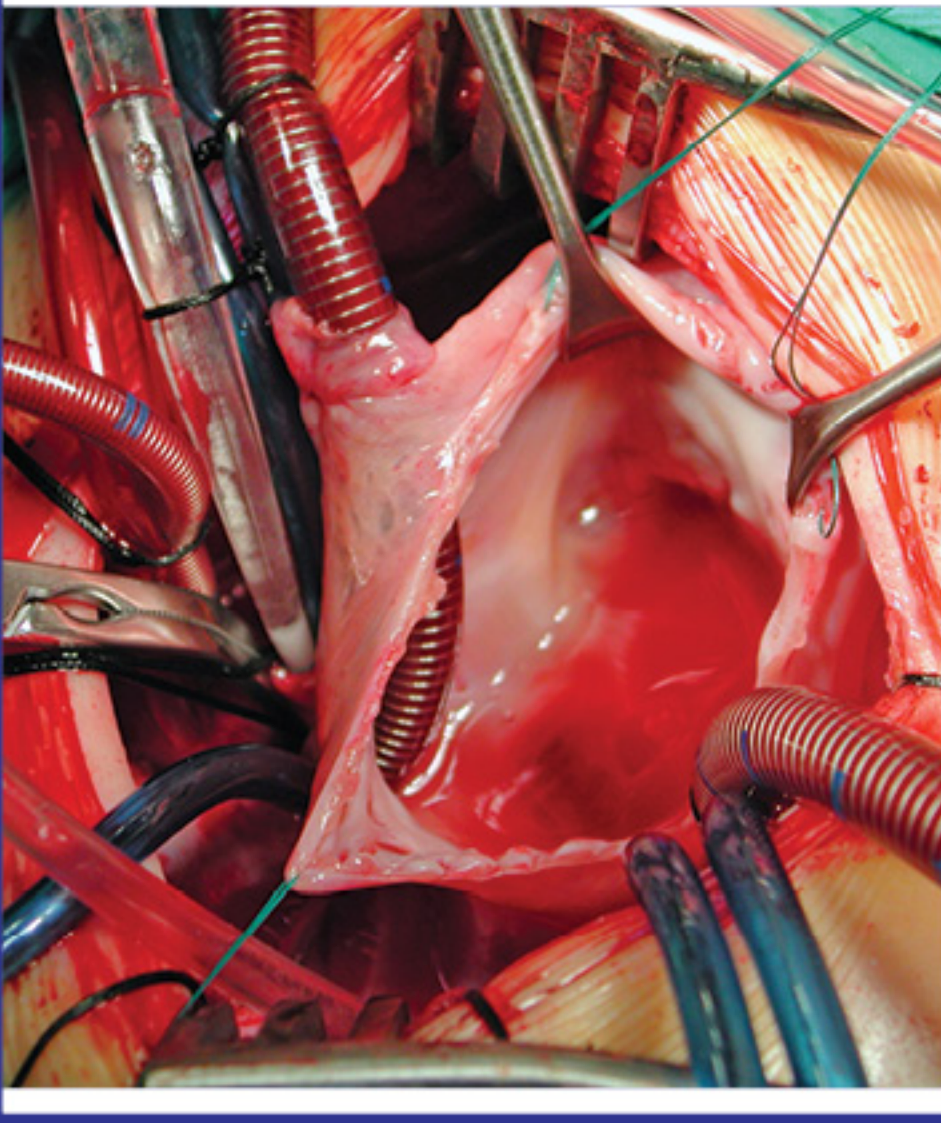
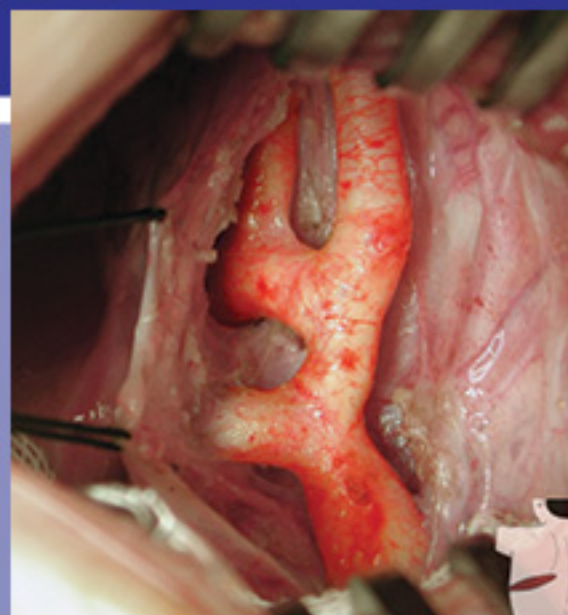
A. SUKRU MERCAN • ZAKARIYA HUBAIL • S. YEN HO

Congenital Heart Disease

A Surgical Color Atlas





Foreword by **HISASHI NIKAIDOH**

cardiotext

CONGENITAL HEART DISEASE

A SURGICAL COLOR ATLAS

CONGENITAL HEART DISEASE

A SURGICAL COLOR ATLAS

AUTHORS

A. Sukru Mercan, MD, FETCS
Consultant Pediatric Cardiac Surgeon
Head, Pediatric Cardiovascular Services
Saad Specialist Hospital
Al Khobar, Saudi Arabia

Zakariya Hubail, BMedSc, MD, FAAP
Consultant Pediatric Cardiologist
Salmaniya Medical Complex, and
Arabian Gulf University
Manama, Bahrain

S. Yen Ho, PhD, FRCPath, FESC, FHEA
Professor of Cardiac Morphology
Consultant, Royal Brompton Hospital &
Harefield NHS Foundation Trust
Imperial College London
London, UK

cardiotext PUBLISHING
Minneapolis, Minnesota

Cardiotext Publishing, LLC
3405 W. 44th Street
Minneapolis, Minnesota 55410
USA

www.cardiotextpublishing.com

Any updates to this book may be found at: www.cardiotextpublishing.com/congenital-heart-disease-a-surgical-color-atlas/

Comments, inquiries, and requests for bulk sales can be directed to the publisher at: info@cardiotextpublishing.com.

Library of Congress Control Number: 2014957046

ISBN: 978-1-935395-90-4

eISBN: 978-1-935395-32-4

Printed in the United States of America

CONTENTS

ABOUT THE AUTHORS

A. Sukru Mercan, MD, FETCS, Consultant Pediatric Cardiac Surgeon, Head of Pediatric Cardiovascular Services, Saad Specialist Hospital, Al Khobar, Saudi Arabia

Dr. Mercan has operated on more than 3,000 cases in the field of congenital cardiac cardiology since 1996. He has established new pediatric cardiac centers in Istanbul, Turkey (2004), and in Al Khobar, Saudi Arabia (2007). He has been actively involved with digital photography and the capturing of intraoperative images of cardiac anomalies for the last 15 years. He is also the Deputy Chief of Cardiovascular Services at Saad Specialist Hospital.

Zakariya Hubail, BMedSc, MD, FAAP, Consultant Pediatric Cardiologist, Salmaniya Medical Complex and Arabian Gulf University, Manama, Bahrain

Dr. Hubail has trained and worked in Bahrain, the United States, and Saudi Arabia. He is currently a consultant pediatric cardiologist at the Salmaniya Medical Complex in Bahrain, and also holds a teaching post at the Department of Pediatrics, College of Medicine, Arabian Gulf University. He has published in local and international journals, and is also a referee for many local journals.

S. Yen Ho, PhD, FRCPath, FESC, FHEA, Professor of Cardiac Morphology, Consultant, Royal Brompton Hospital & Harefield NHS Foundation Trust, Imperial College London, London, UK

Professor Ho carries out research on the morphology and anatomy of normally structured hearts, hearts with conduction disturbance, and hearts with congenital malformations. She conducts regular courses on morphology of congenital heart defects and on cardiac anatomy for surgeons and interventionists.

FOREWORD

Modern medicine dawned with *De Humani Corporis Fabrica*, published in 1543 by Andreas Vesalius. The accurate anatomy based on direct observation began the new era of evidence-based medicine. Although Vesalius clearly described the anatomy of the human heart, it took nearly another century before the function of the heart was clarified by William Harvey in *De Motu Cordis* in 1628.

More recently, in the past 2 centuries, modern surgeons expanded their territory from the abdominal cavity to the thoracic and intracranial spaces and, finally, the inside of the human heart. The accurate knowledge of anatomy and the clear understanding of cardiovascular and pulmonary functions brought about the golden era of cardiovascular surgery and interventional cardiology practiced by medical professionals today.

In 1966, I was introduced to pediatric cardiac surgery at Children's Memorial Hospital (now Lurie Children's Hospital) in Chicago. The team, which was gathered by Dr. Willis J. Potts (1895–1963), was in full function, and I worked closely with Dr. Farouk S. Idriss (1928–1992), who was a pioneer in congenital heart surgery. Dr. Idriss shared an interest in the anatomy of congenital heart disease with Dr. Maurice Lev (1908–1994) of the Hektoen Institute in Chicago. Dr. Lev contributed immensely, as a pathologist, in the understanding of congenital heart disease.

Dr. Idriss began collecting surgical photographs of congenital heart defects, while I enjoyed my hobby of making line drawings of operative findings. We talked about making an album of these photographs with illustrations from a surgeon's point of view. After acquiring hundreds of photographs, I left Chicago at the end of 1977, and the project was abandoned. The need for such a book of anatomy for congenital heart defects remained real to this day.

This need and my dream are realized in this *Atlas* by the impressive work of 3 doctors: a surgeon, a cardiologist, and a pathologist. This *Atlas* includes a remarkably wide range of congenital heart disease with details of anatomy and clinical information. The beautiful pictures teach us much about surgical repair over and above the pathological anatomy. Understandably, congenital heart disease includes numerous variations, and even this *Atlas* cannot give totally comprehensive coverage. Yet, this *Atlas* may very well be the most accurate photographic description of congenital heart disease ever published.

We, the community of medical professionals, should be grateful to the authors, as I believe that this *Atlas* will help countless infants, children, and young adults by guiding their doctors, both surgeons and cardiologists, toward better understanding of the anatomic details of their heart defects.

The more we learn in medicine, the more we are amazed how "fearfully and wonderfully our bodies are made" (Psalm 139:14).

Hisashi Nikaidoh, MD, FACS, FAAP
Professor of Cardiothoracic Surgery (Retired)
University of Texas Southwestern Medical Center
Dallas, Texas

PREFACE

Congenital heart disease constitutes an important cause for morbidity and mortality in pediatrics. Interventional cardiology has made important strides over the past 2 decades, but despite this, many congenital malformations still require surgical treatment. Recent advances have made it possible to repair or palliate almost all types of congenital heart defects. This book is a pictorial illustration of congenital heart disease. The pictures in this *Atlas* represent real views as seen through the surgeon's eyes, from actual patients in the operating room. The old saying that "a picture is worth a thousand words" cannot be overemphasized.

The material was collected by one of the authors intraoperatively using a digital camera. The images were processed by computer software, and line drawings were made to clarify the anatomic and surgical details. Numbers and legends were used to mark areas of interest. We avoided the overuse of arrows because they may obscure views and, when numerous, make the pictures look overcrowded. The numbering starts from the top of the figure and continues in a clockwise direction. A brief clinical history is provided to keep the surgical procedure in context with the clinical status. Important surgical, anatomic, and pathologic points are highlighted. However, we kept the text to a minimum because this is primarily an atlas.

This book is intended for general pediatric surgeons, cardiac surgeons, pediatric cardiologists, cardiac pathologists, cardiac intensivists, and those interested in congenital heart disease. We hope this *Atlas* will complement the already existing textbooks of pediatric cardiology and cardiac surgery.

We are grateful to our patients, who are the material of this book; to our colleagues for help and encouragement; and to our families for their support and the time "sabbatical" they granted us to prepare this book.

A.S. Mercan
Z. Hubail
S. Yen Ho
2015

ACKNOWLEDGMENT

We would like to acknowledge the contribution of Professor Mehmet Salih Bilal, Istanbul, Turkey, and Professor Emin Tireli, Istanbul University, Istanbul, Turkey, who shared with us some of the photographs of their patients. Professor Bilal has graciously provided us with the following photographs.

Pulmonary Atresia: Figures 14.8 to 14.11, 14.14 to 14.23

Congenitally Corrected Transposition of the Great Arteries: Figures 16.24 to 16.39

Single Ventricle: Figures 17.7 to 17.14, 17.38 to 17.40

Aortic Valve: Figures 19.26 to 19.42

Valve Surgery: Figures 20.24 to 20.28

Coronary Artery Abnormalities: Figures 21.19 to 21.28

Professor Emin Tireli has provided us with the following photographs.

Transposition of the Great Arteries: Figures 15.23 to 15.37

ABBREVIATIONS

3D	three-dimensional
ALCAPA	anomalous left coronary artery from the pulmonary artery
APW	aortopulmonary window
ASD	atrial septal defect
AV	atrioventricular
AVSD	atrioventricular septal defect
BT shunt	Blalock-Taussig shunt
ccTGA	congenitally corrected transposition of the great arteries
CPB	cardiopulmonary bypass
CS	coronary sinus
DILV	double inlet left ventricle
DKS	Damus-Kaye-Stansel
DORV	double outlet right ventricle
d-TGA	*(see TGA)*
ePTFE	expanded polytetrafluoroethylene
HLHS	hypoplastic left heart syndrome
ICU	intensive care unit
IVC	inferior vena cava
LAA	left atrial appendage
LAD	left anterior descending (coronary artery)
l-TGA	L-transposition of the great arteries
LV	left ventricle; left ventricular
LVOT	left ventricular outflow tract
MAPCA	major aortopulmonary collateral artery
MPA	main pulmonary artery
PA	pulmonary artery
PDA	patent ductus arteriosus
PFO	patent foramen ovale
PTFE	polytetrafluoroethylene
PV	pulmonary vein
RAA	right atrial appendage
RV	right ventricle; right ventricular
RVOT	right ventricular outflow tract
SVC	superior vena cava
TAPVC	totally anomalous pulmonary venous connection
TAPVD	totally anomalous pulmonary venous drainage
TGA	transposition of the great arteries
TOF	tetralogy of Fallot
VATS	video-assisted thoracoscopic surgery
VSD	ventricular septal defect

CHAPTER 1

Introduction

Cardiac surgery effectively started with Robert Gross closing a patent ductus arteriosus (PDA) in 1938.[1] The next major advance was palliation of cyanotic heart disease by systemic-to-pulmonary shunt, performed by Alfred Blalock in 1944.[2] However, it was the cardiopulmonary bypass machine (1953) that ushered in modern cardiac surgery.[3] It enabled performing intracardiac repair under direct vision in a bloodless field. Coupled with accurate preoperative assessment and sophisticated postoperative intensive care, congenital heart surgery has made, and continues to make, significant strides. Nowadays even the most complex heart diseases can be palliated or repaired with low mortality and acceptable morbidity. In recent years, there has been a trend toward earlier repair for most lesions.

Interventional cardiac catheterization has also advanced significantly, and has changed the way we currently treat many congenital heart diseases. In due time, some of the techniques described in this book will be replaced by catheter-based interventions or hybrid procedures.

Although the outcomes of congenital heart disease have improved significantly, there are still a few lingering issues. With the growing population of survivors of CHD came the realization of important physical, neurodevelopmental, and psychosocial morbidities that may affect the overall quality of life.[4]

STERNOTOMY

A median sternotomy (Figures 1.1–1.10) is the approach most commonly utilized for the majority of congenital heart surgeries.

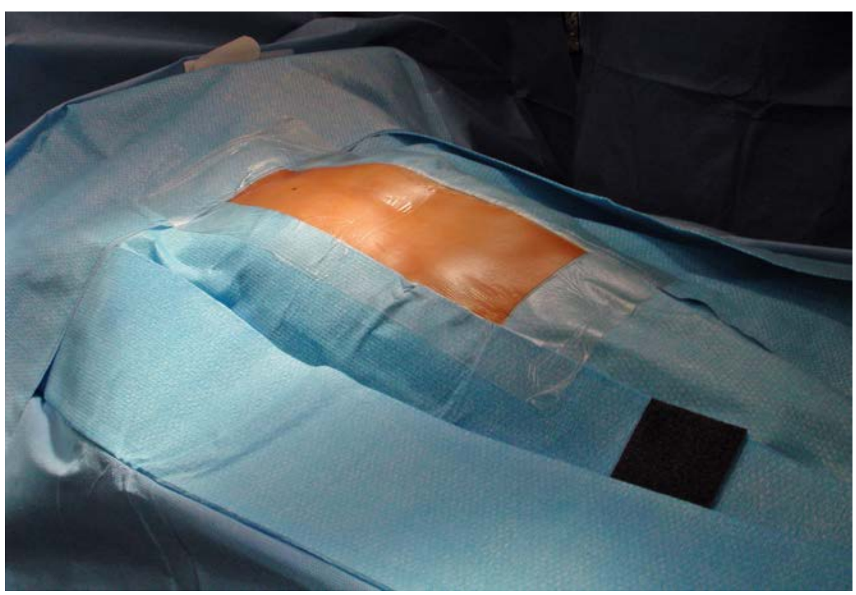


Figure 1.1. A child on the operating table, anesthetized, prepped, and draped, prior to skin incision for repair of congenital heart disease via a median sternotomy.

Figure 1.2. A skin incision has been made for a standard median sternotomy.

Upon opening the sternum, the anterior mediastinal structures come into view. The thymus is large in children and begins to involute at puberty. It is usually sub-totally excised to facilitate access (Figures 1.4–1.6). The heart and the proximal great vessels are wrapped by the pericardial sac. Cardiopulmonary bypass (CPB) is established by cannulating the superior vena cava (SVC) and inferior vena cava (IVC). The blood is oxygenated in the CPB machine and returned back through an arterial cannula to the ascending aorta. A cardioplegia catheter is inserted into the aorta proximal to the site of arterial cannula. Between these 2 catheters, an aortic cross-clamp is placed (Figure 1.7). The operative field is often small and crowded with instruments (Figure 1.8). Metal wires are usually used to close the sternum after the repair.

Occasionally a submammary incision is made, especially in females for cosmetic reasons (Figure 1.11). Through this incision a limited number of lesions, such as atrial septal defects (ASD), can be repaired.

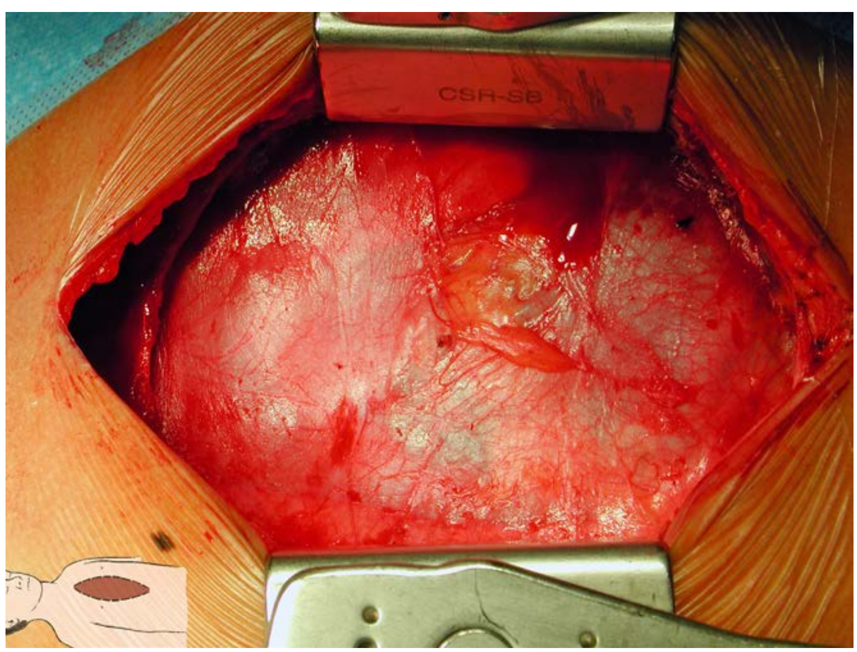


Figure 1.3. The sternal bone has been cut, and a pair of retractors placed. The anterior mediastinal structures are seen.

Figure 1.4. A median sternotomy view from another child undergoing heart surgery. The thymus gland is usually large in children.

1. Thymus gland
2. Pericardial sac

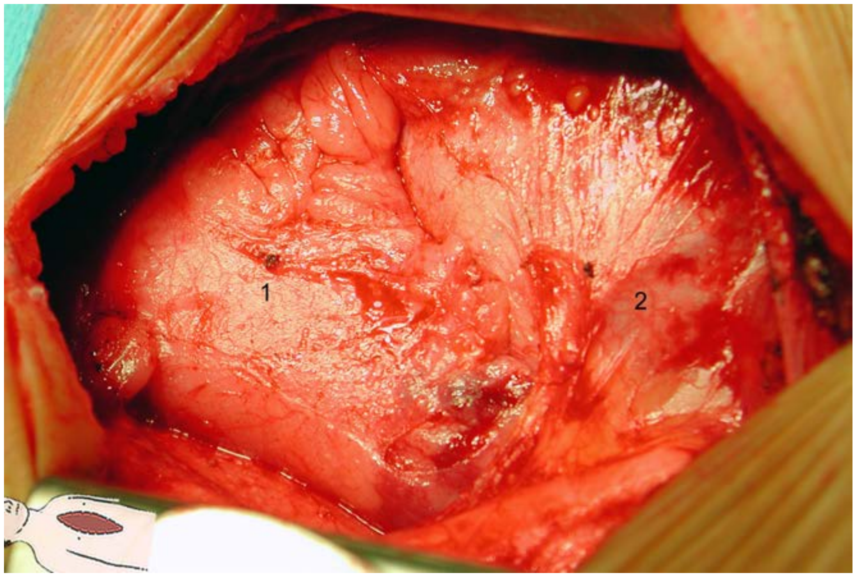


Figure 1.5. The thymus gland is mobilized prior to excision.

1. Left lobe of thymus
2. Right lobe of thymus
3. Heart

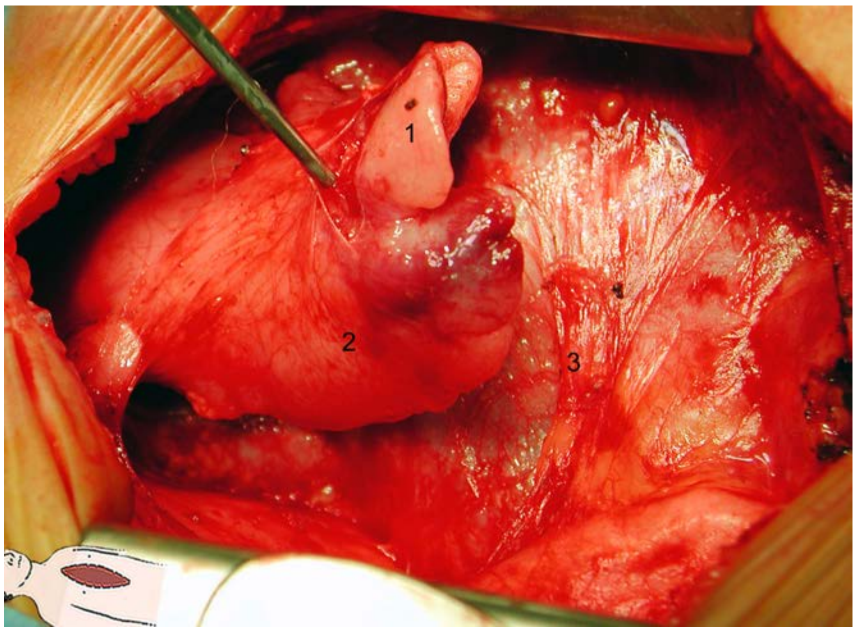
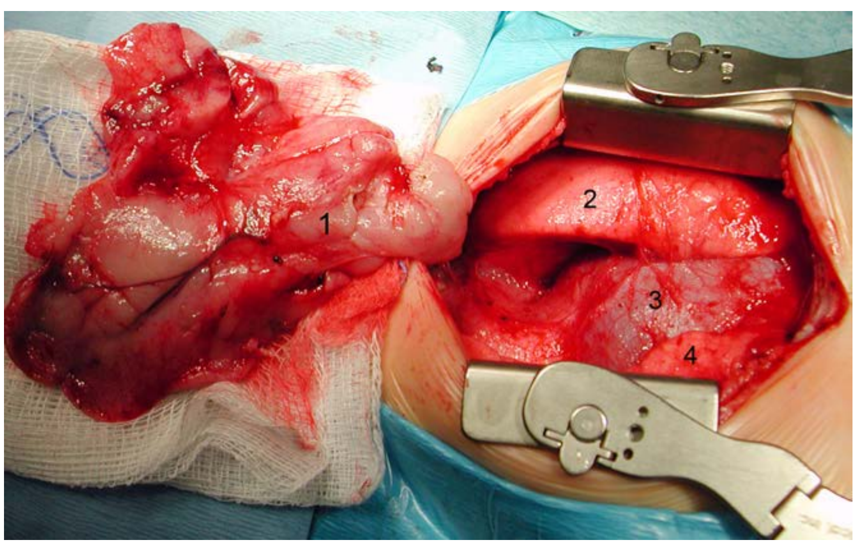


Figure 1.6. The thymus is excised to gain better access to the heart and great vessels for repair and cannulation.

1. Thymus
2. Left lung
3. Heart
4. Right lung



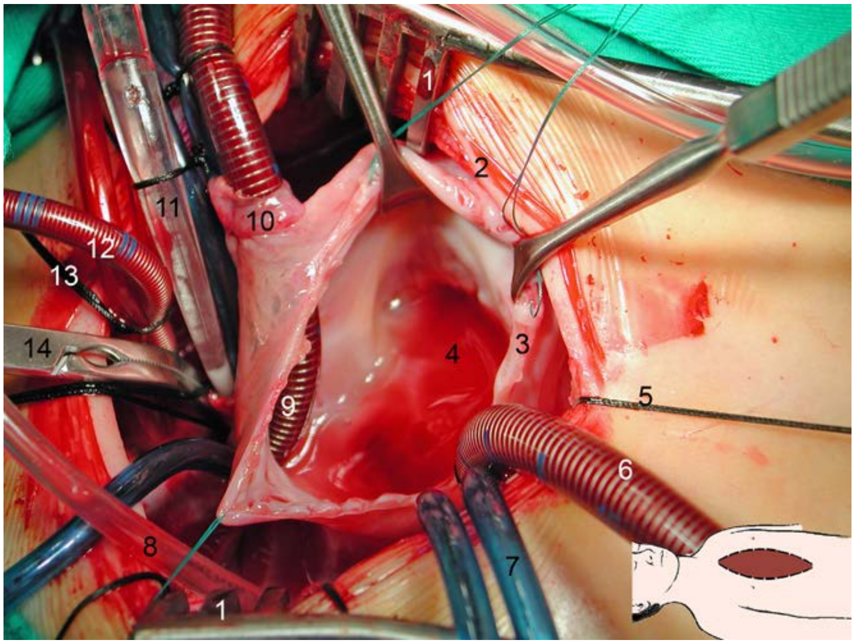
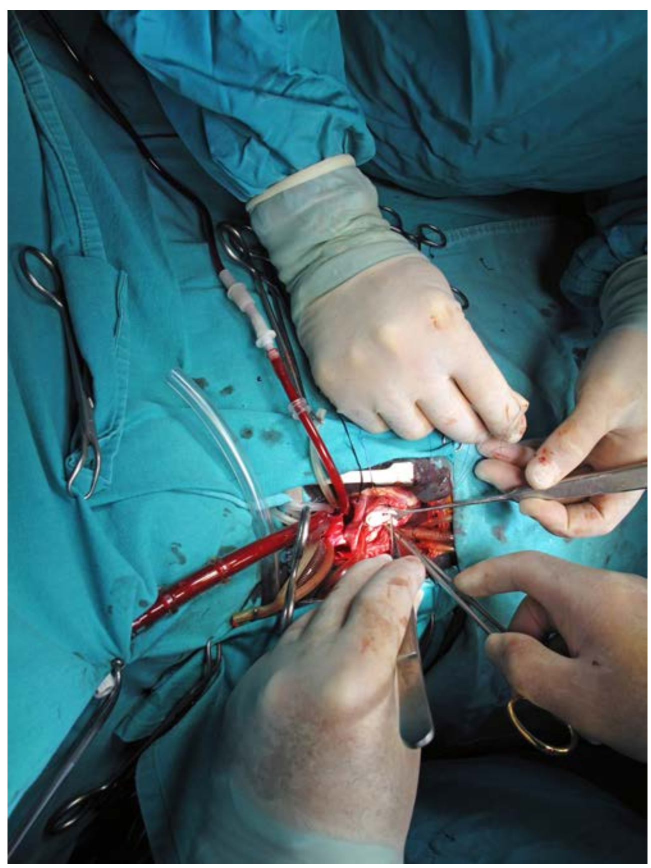


Figure 1.7. The operative field through a median sternotomy and right atriotomy in a 2-year-old child with an ASD.

1. Sternal retractor
2. Skin incision
3. Atriotomy incision
4. Blood in floor of opened right atrium
5. Stay suture for pericardium
6. IVC venous cannula
7. Tourniquet
8. Suction catheter
9. SVC venous cannula
10. Entry of SVC cannula through atrial appendage
11. Cardioplegia catheter
12. Aortic cannula
13. Stay suture isolating ascending aorta
14. Aortic cross-clamp

Figure 1.8. The operative field of another patient, an infant, undergoing cardiac surgery. The surgeons have to work through a small window cluttered with cannulas, catheters, tourniquets, retractors, and surgical instruments. Magnifying loops are indispensable for good visualization.



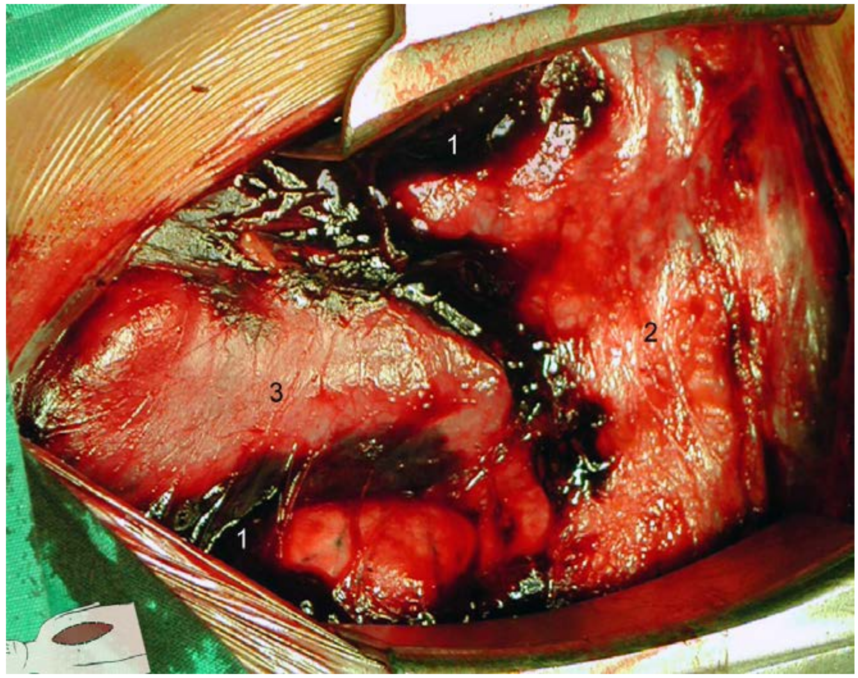


Figure 1.9. Median sternotomy view of a child undergoing heart surgery. Clotted blood is seen around the heart and thymus. It came from a vein inadvertently perforated during placement of a central venous catheter by the anesthesiologist.

1. Clotted blood
2. Heart
3. Thymus

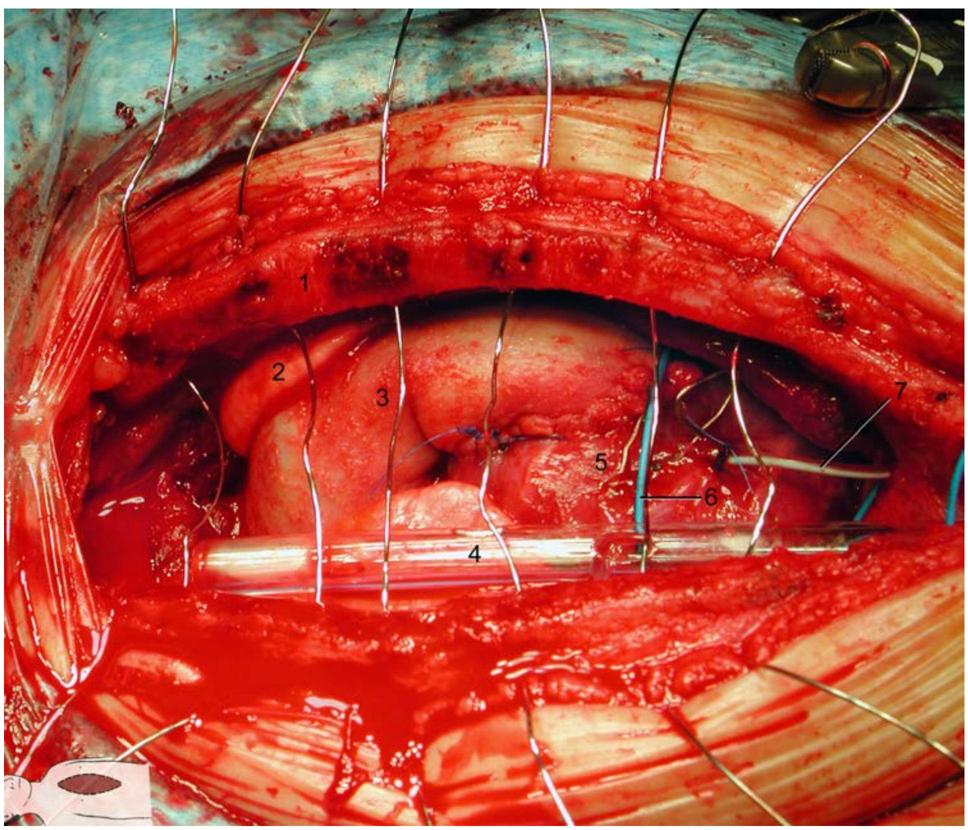


Figure 1.10. A median sternotomy view at the conclusion of a heart surgery, just prior to sternal closure. The patient, a 4-month-old infant with double outlet left ventricle (LV), underwent ventricular septal defect (VSD) closure and right ventricle–to–pulmonary artery (RV-to-PA) conduit implantation.

1. Sternum bone
2. Left lung
3. RV-to-PA conduit (Contegra)
4. Mediastinal chest tube
5. RV
6. Temporary pacing wire
7. Pulmonary artery catheter