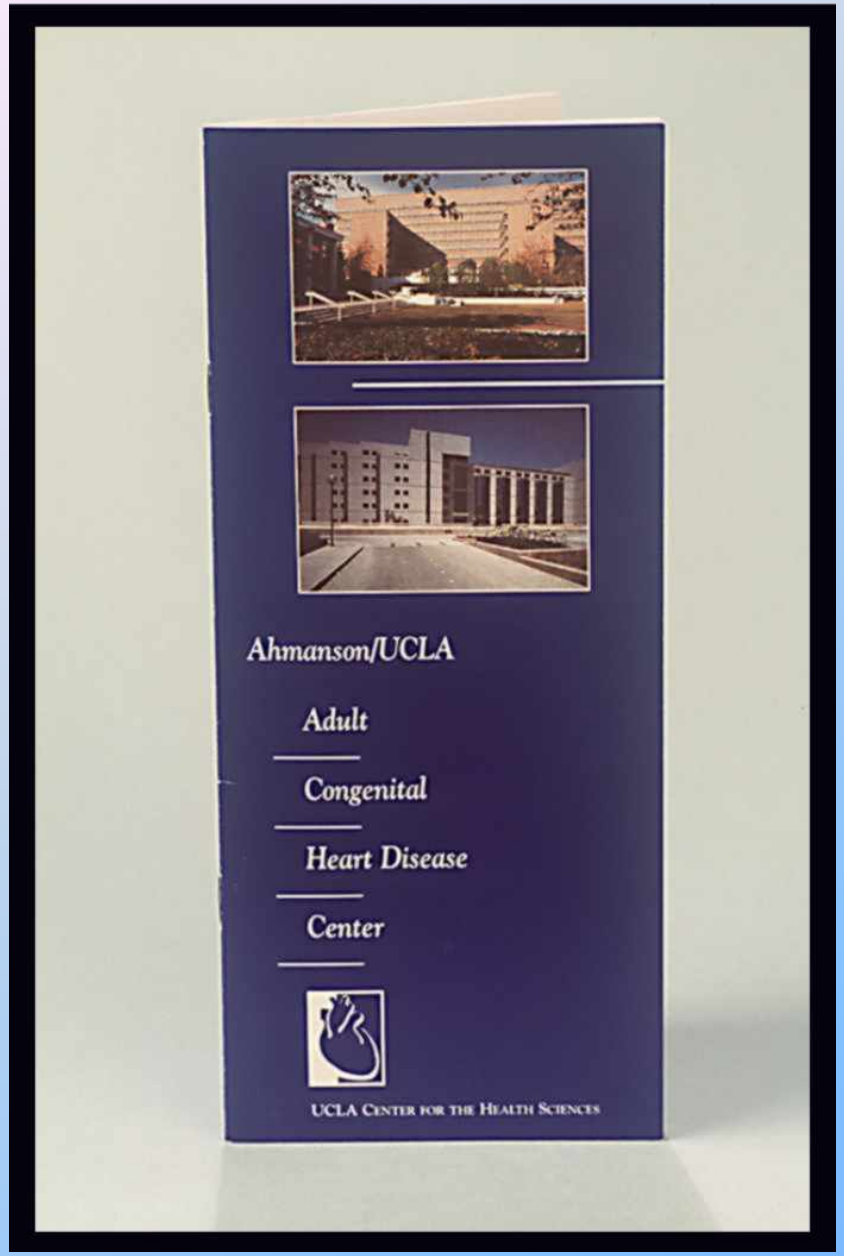


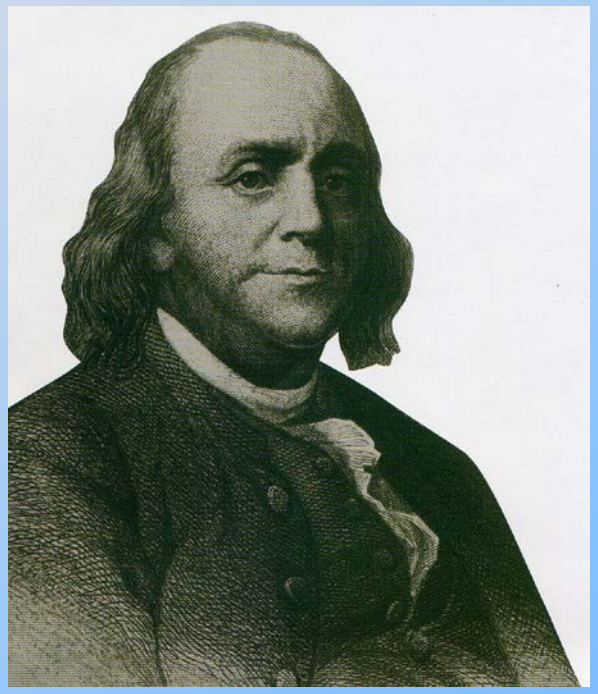
Adult Congenital Heart Disease

Historical Perspectives and Personal Reminiscences

Joseph K. Perloff

Children Are Not Small Adults.
Adults Are Not Large Children.

IN THE YEAR OF CHRIST
MDCCLV.
GEORGE THE SECOND HAPPILY REIGNING
(FOR HE SOUGHT THE HAPPINESS OF HIS PEOPLE)
PHILADELPHIA FLOURISHING
(FOR ITS INHABITANTS WERE PUBLICK SPIRITED)
THIS BUILDING
BY THE BOUNTY OF THE GOVERNMENT.
AND OF MANY PRIVATE PERSONS,
WAS PIOUSLY FOUNDED
FOR THE RELIEF OF THE SICK AND MISERABLE:
MAY THE GOD OF MERCIES
BLESS THIS UNDERTAKING.



First Hospital in America

Pennsylvania Hospital

1751

The Childrens' Hospitals

The Hospital for Sick Children established in London in 1852 with the aid of Charles Dickens was the first major medical facility in the English-speaking world dedicated to treatment of the young. The second major facility, Children's Hospital of Philadelphia, was founded three years later, and Children's Hospital of Boston opened in 1869. Until the turn of the 20th century, these institutions were little more than dim lights of hope in the darkness of pediatric medicine.

Where Was Congenital Heart Disease ?

Osler's THE PRINCIPLES AND PRACTICE OF MEDICINE

1892

The Section on Diseases of the Circulatory System contained a five page chapter devoted to *Congenital Affections of the Heart*.

CONGENITAL AFFECTIONS OF THE HEART

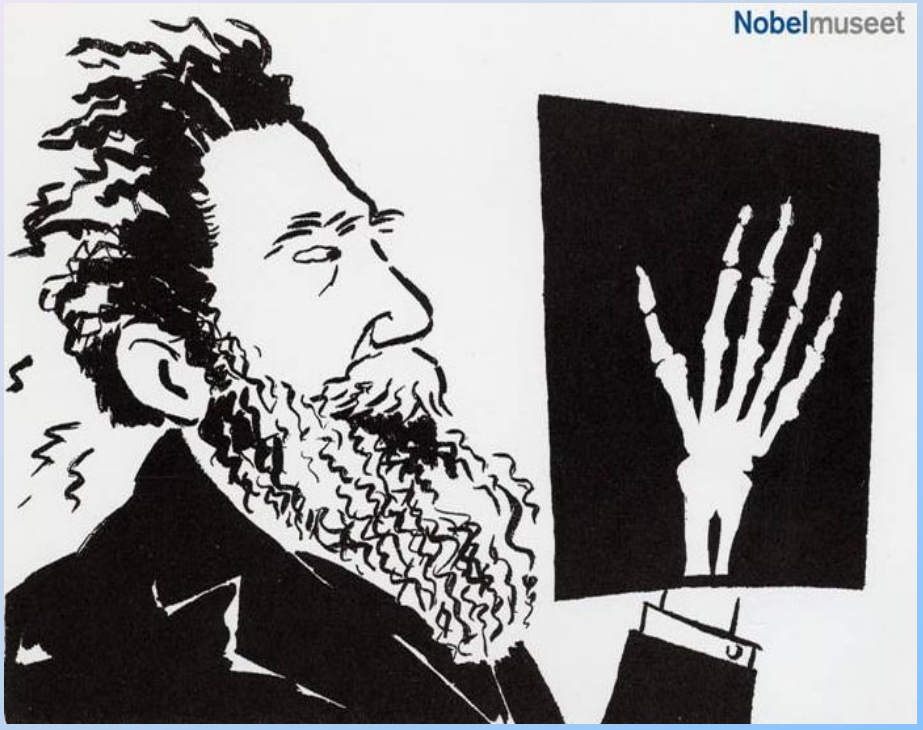
These have only a limited clinical interest, as in a large proportion of the cases the anomaly is not compatible with life, and in others nothing can be done to remedy the defect or even to relieve the symptoms.

Holt's THE DISEASES OF INFANCY AND CHILDHOOD

1897

The Section on Diseases of the Circulatory System contained a seven page chapter devoted to *Congenital Anomalies of the Heart.*

1895



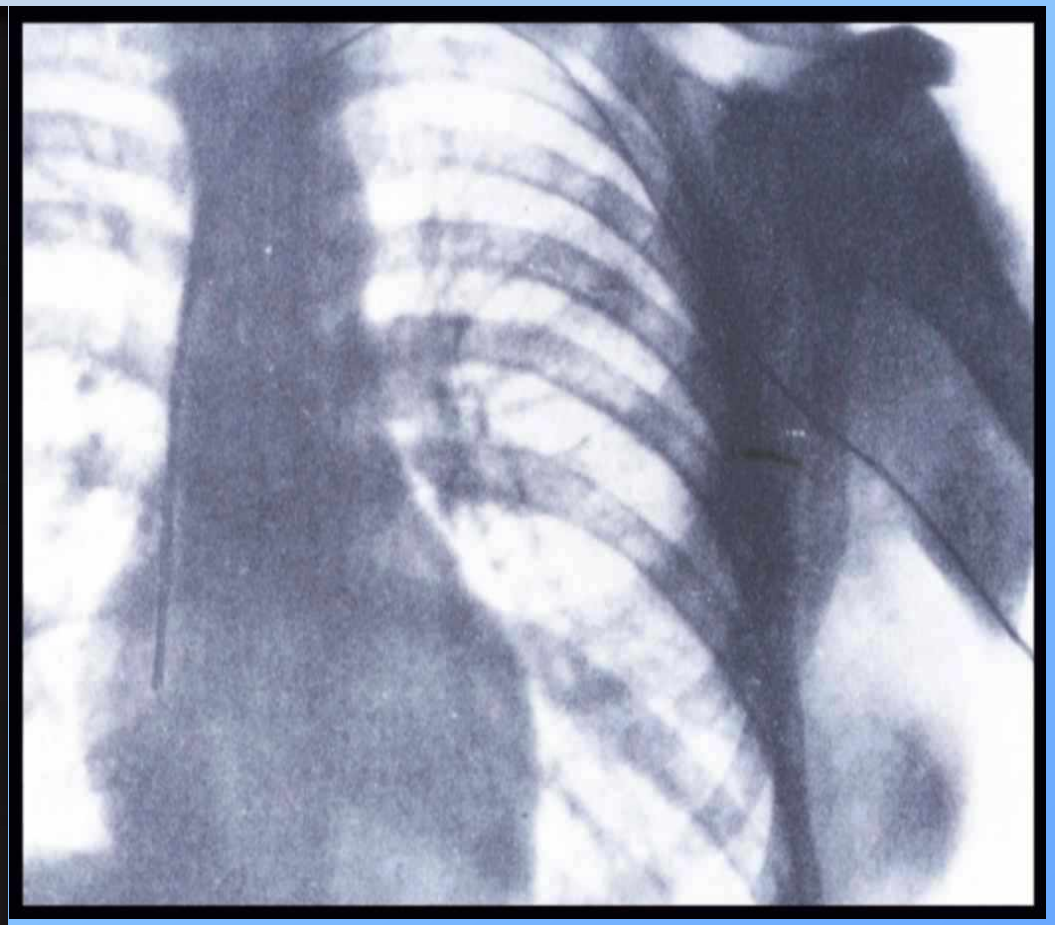
When he held his hand before the fluorescing screen, he was astonished to see the bones of his thumb and fingers. Wilhelm Konrad Roentgen, a German experimental physicist, was awarded the first Nobel prize in 1901.

The Internal Structure of the Living Heart

Four decades after Roentgen's discovery of "a new kind of rays," angiography had its inception with injection of contrast materials into blood vessels of cadavers and animals.

In 1937 Castellanos, Pereiras, and Garcia in Havana visualized the right cardiac chambers in infants and children. George Potts Robb and Israel Steinberg in New York, developed angiography in adults. "*The internal structure of the living heart had been revealed for the first time…*"

Werner Forssman
The Cardiac Catheter



1929

Prelude to the Nobel

Measurement of cardiac output used the principle described by Fick and Grollman in 1870. Andre F. Cournand and Dickenson W. Richards of New York had been catheterizing dogs since 1937. Forssmann's cardiac catheter provided a means of applying the Fick principle in human subjects.

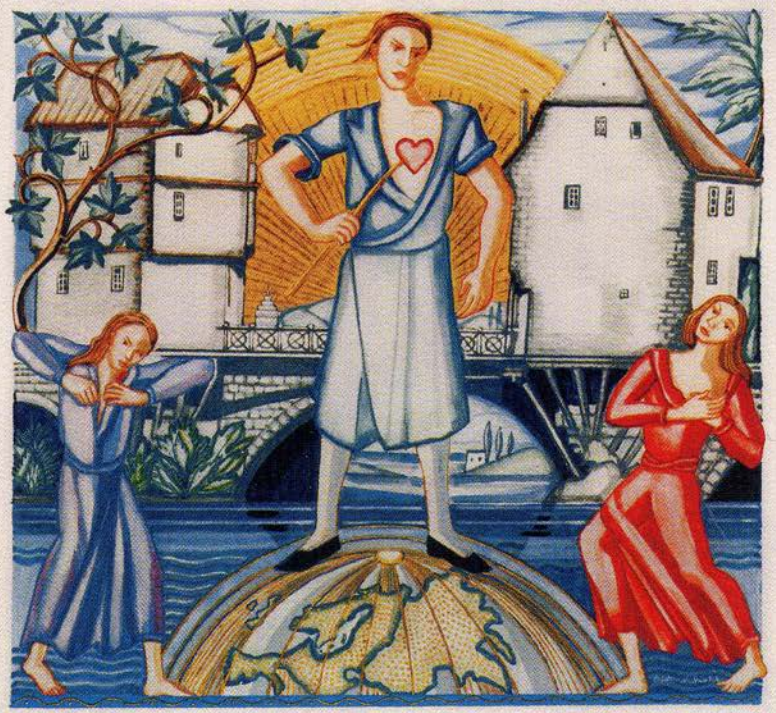
The Nobel Prize in Physiology or Medicine 1956. "For their discoveries concerning heart catheterization and pathological changes in the circulatory system"

Andre Frederic Cournand

Werner Forssmann

Dickinson W. Richards

Karolinska Institute Stockholm



KUNGL. KAROLINSKA
MEDIKO-KIRURGISKA
INSTITUTET

VILKET ENLIGT TESTAMENTE SOM
DEN 27 NOVEMBER 1895 UPPRÄTTATS AV

ALFRED NOBEL

ÄGER ATT MED NOBELPRIS BELÖNA
DEN VIKTIGASTE UPPTÄCKT VARMED DE

FYSIOLOGISKA OCH MEDICINSKA VETEN-
SKAPERNA UNDER SENASTE TIDEN RIKTATS
HAR DENNA DAG BESLUTIT ATT TILLERKÄNNA
DET ÅR 1956 UTGÅENDE PRISET ÅT

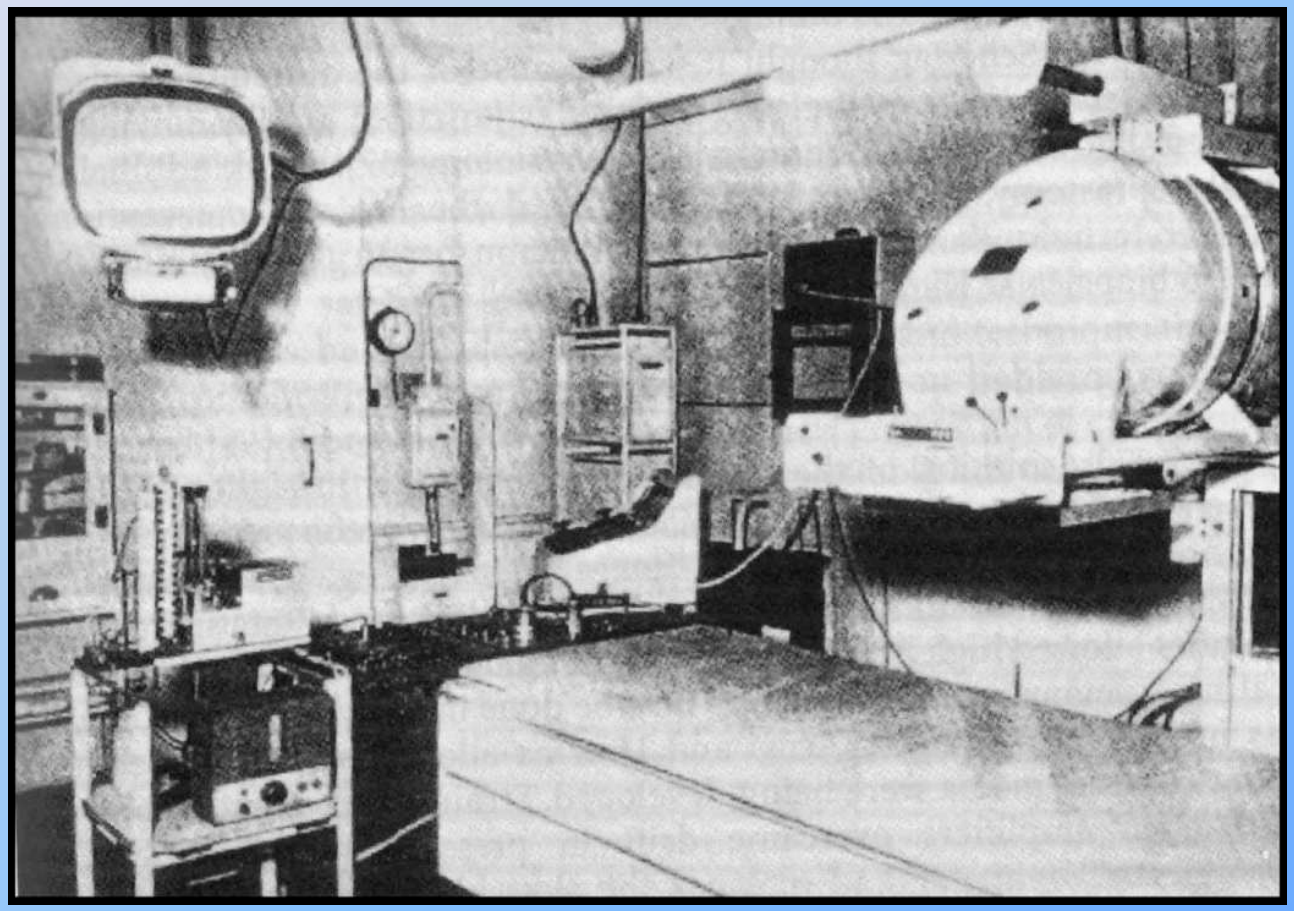
ANDRÉ COURNAND
WERNER FORSSMANN
OCH
DICKINSON W. RICHARDS

GEMENSAMT FÖR DERAS UPPTÄCKTER RÖ-
RANDE HJÄRTKATETRISERING OCH SJUKLIGA
FÖRÄNDRINGAR I CIRKULATIONSAPPARATEN.

STOCKHOLM DEN 18 OKTOBER 1956

Cardiac Catheterization Laboratory

National Heart Hospital, London 1955



Electrical Properties of the Heart

Anatomic 1906

Electrocardiographic 1924

Electrophysiologic 1949

Anatomic 1906

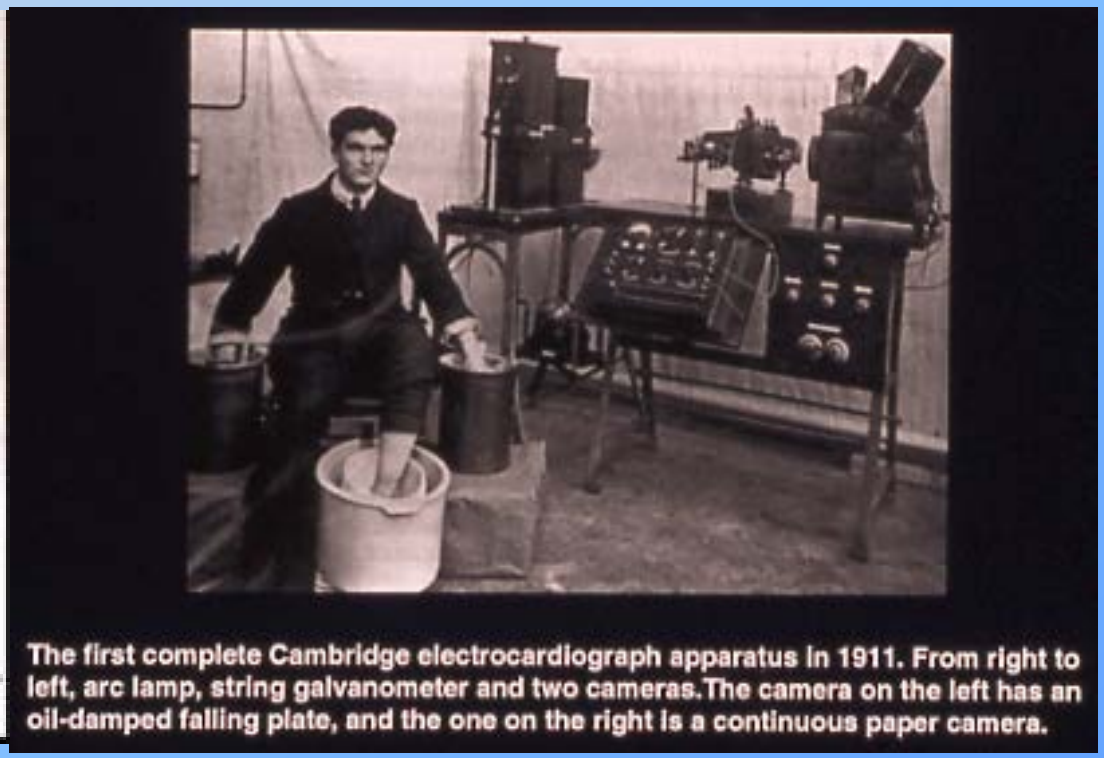
The Conduction System of the Mammalian Heart

In 1906, Suano Tawara working in Ludwig Aschoff's laboratory in Marburg, published his epoch-making monograph.

Electrocardiographic 1924

Willem Einthoven (1860-1927)

Nobel prize in physiology or medicine 1924

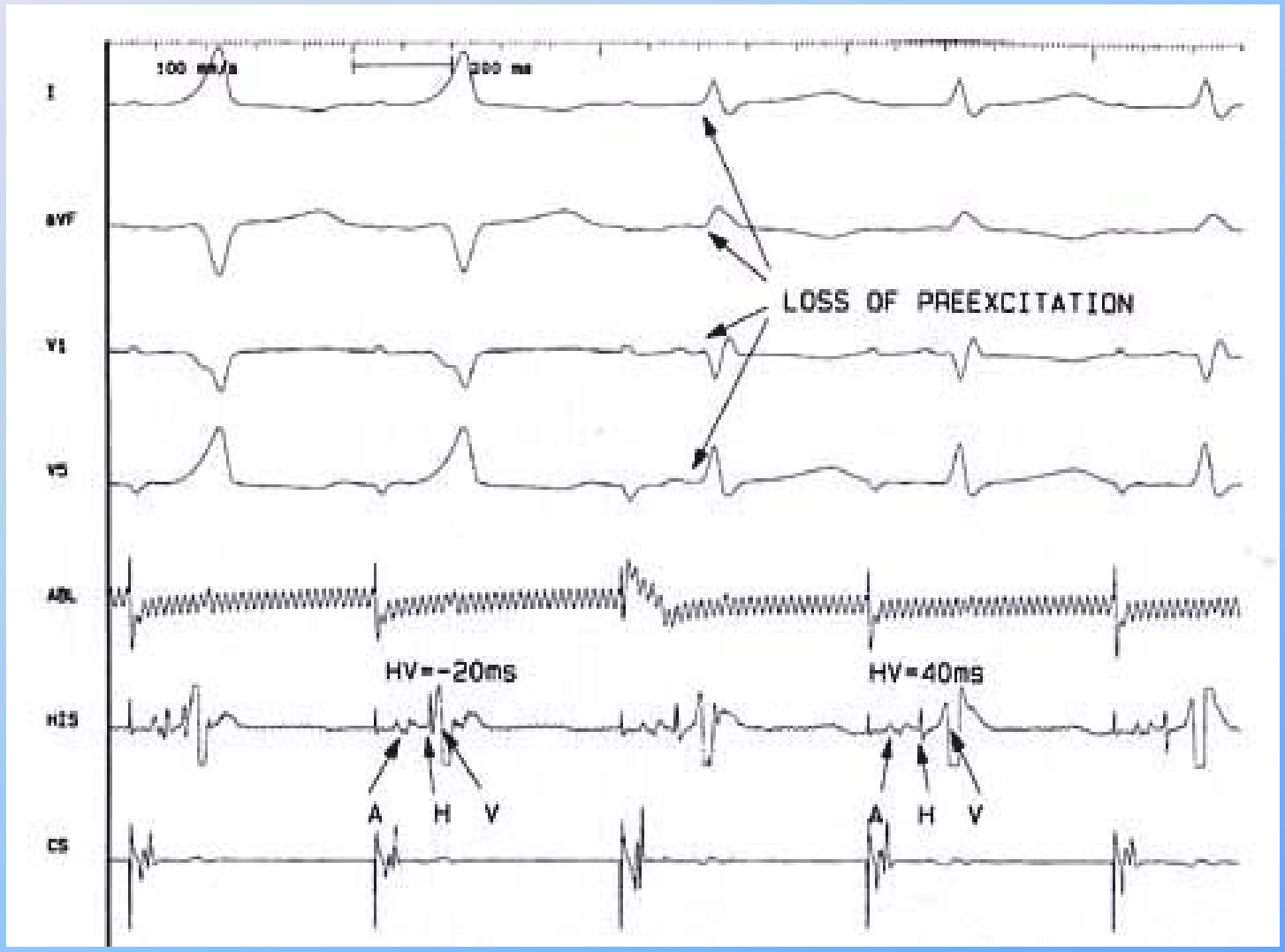


The first complete Cambridge electrocardiograph apparatus in 1911. From right to left, arc lamp, string galvanometer and two cameras.The camera on the left has an oil-damped falling plate, and the one on the right is a continuous paper camera.

Electrophysiologic 1949

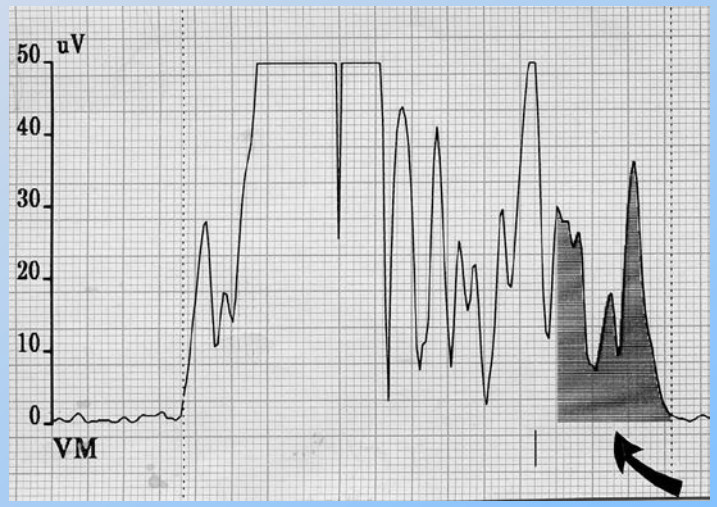
The inception of electrophysiology was in 1949 with the introduction of the microelectrode by Ling and Gerard

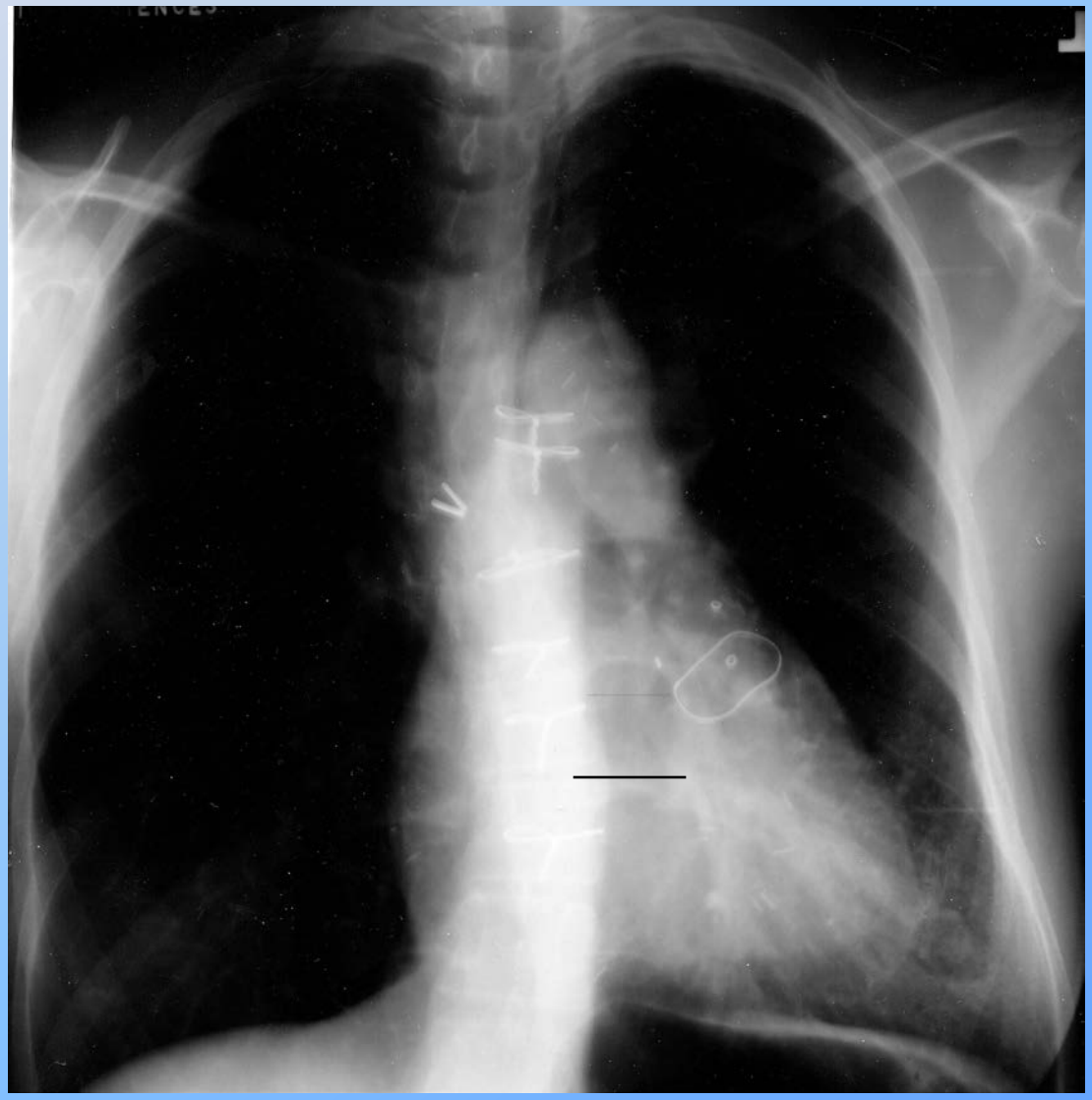
Accessory Pathways Radiofrequency Ablation



A Slow Conduction Substrate

The Signal Averaged Electrocardiogram

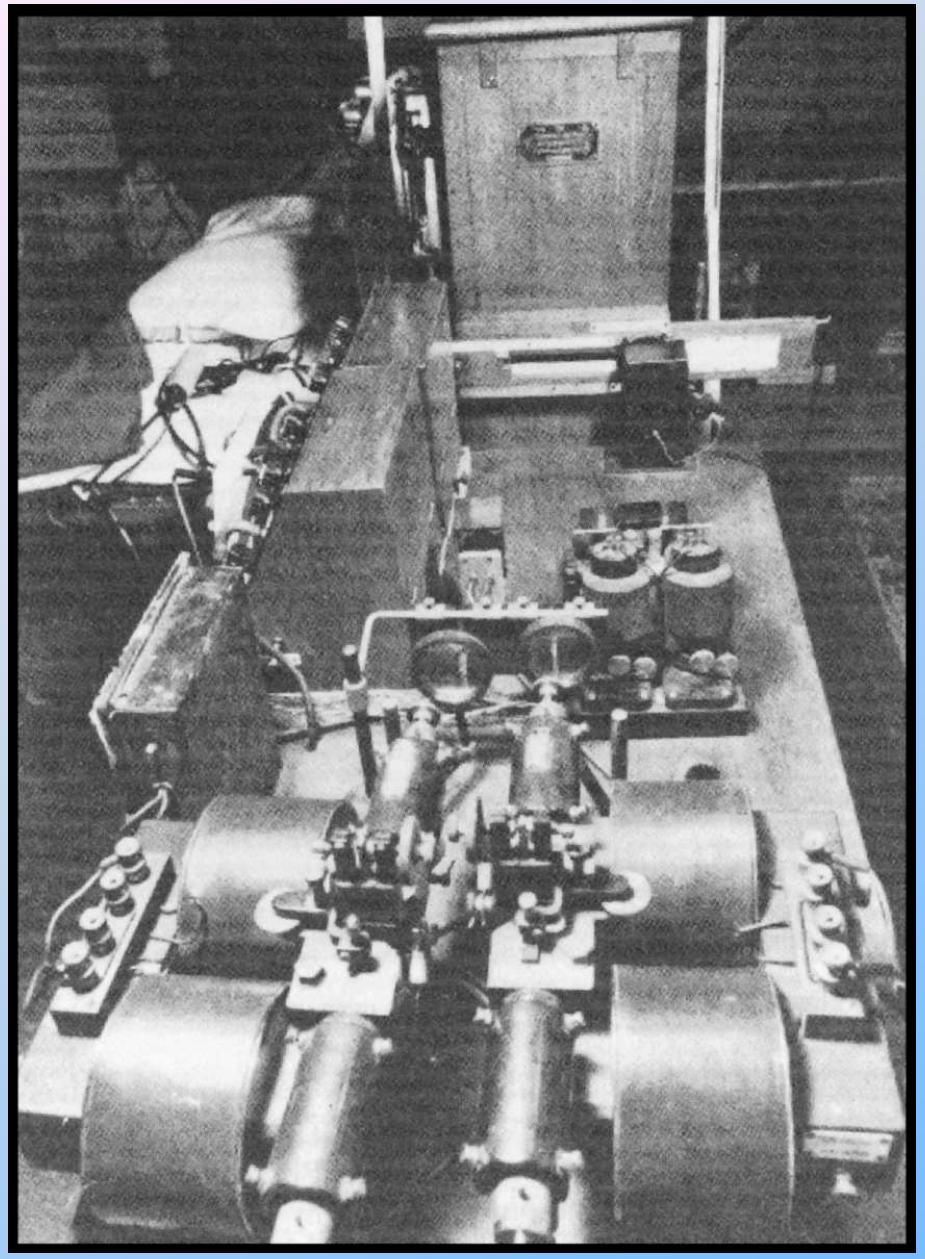




The Audible Language of the Heart

PHONOCARDIOGRAPHY

Willem Einthoven, inventor of the electrocardiogram, was also the first to record heart sounds--the audible language of the heart (1907).

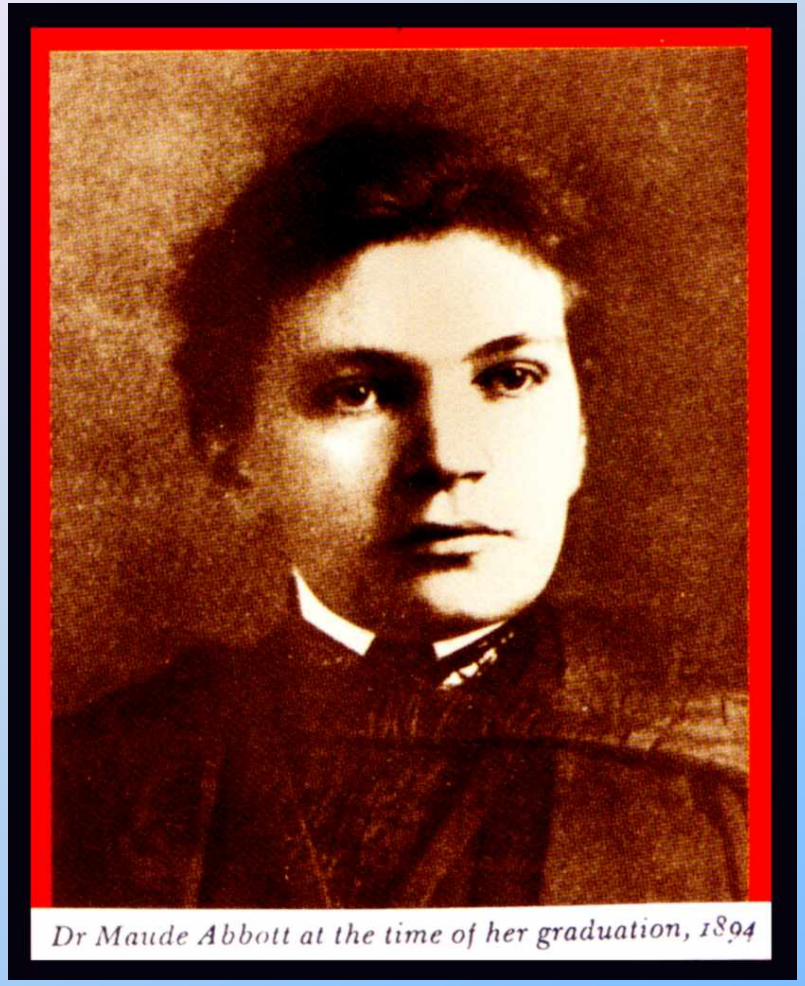


Aubrey Leatham's high-definition phonocardiograph consisting of two Cambridge string and two mirror galvanometers. (Welcome Museum, London.)

Before World War II, congenital malformations of the heart were regarded as *hopeless futilities*, interests appropriate for the few women in medicine. Maude Abbott was advised by William Osler to occupy herself with the collection of anatomic specimens at McGill, and Helen Taussig was advised to occupy herself with the *hopeless futilities* in the children's clinic at Hopkins. Congenital heart disease in adults was an oxymoron.

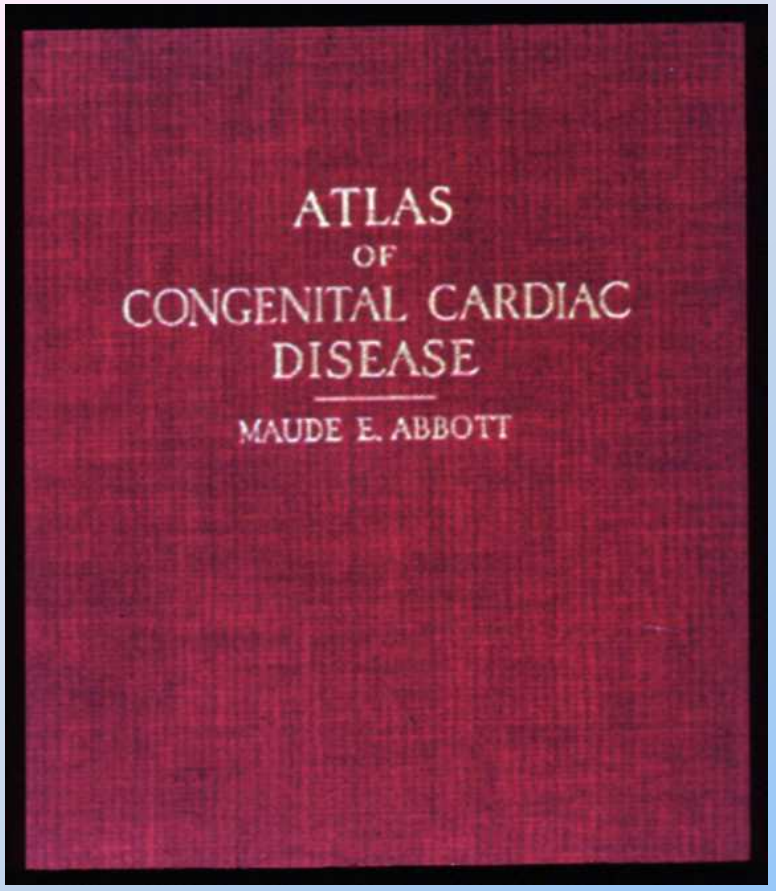
But now, approximately 85% of infants with congenital heart disease in developed countries reach adulthood.

The hopeless futilities have come of age.



Maude Abbott

Helen Taussig



1936

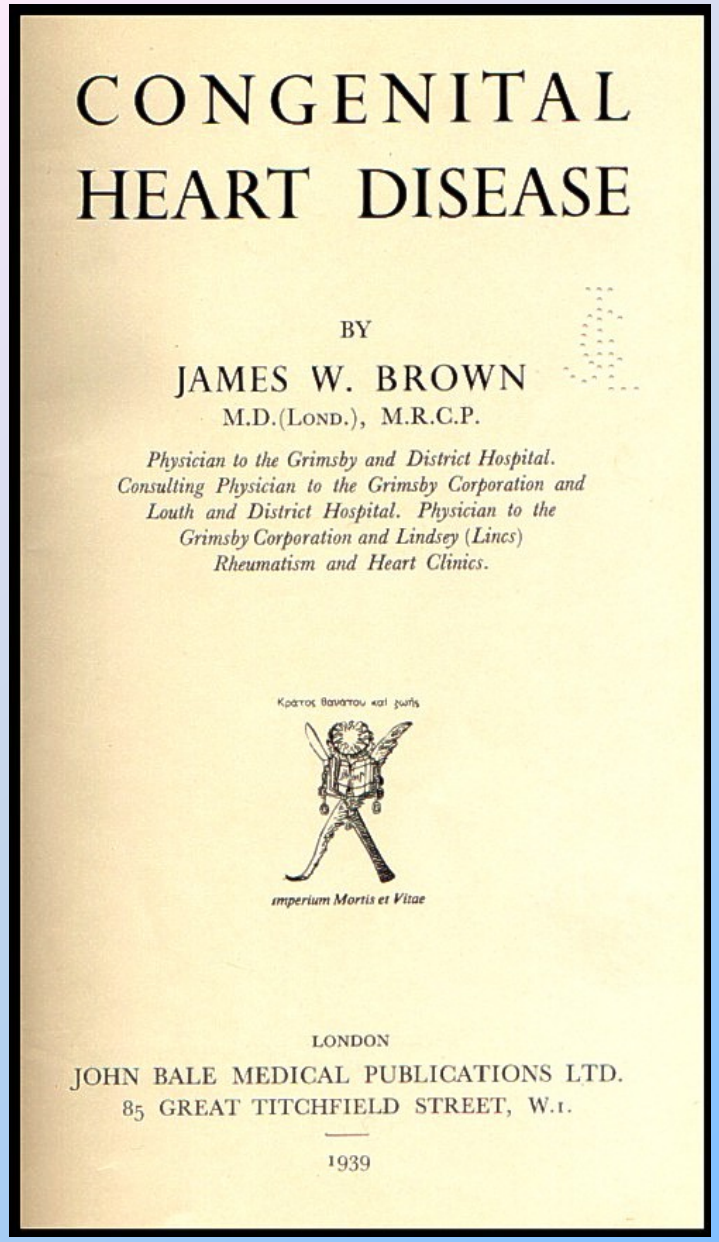
The Osler Library
McGill

The Coming of Age

I'll now focus on two topics:

1) How the coming of age was achieved.

2) Why there are compelling ethical and practical reasons to assume responsibility for the patients who come of age.


CONGENITAL
HEART DISEASE

BY

JAMES W. BROWN

M.D.(LOND.), M.R.C.P.

Physician to the Grimsby and District Hospital. Consulting Physician to the Grimsby Corporation and Louth and District Hospital. Physician to the Grimsby Corporation and Lindsey (Lincs) Rheumatism and Heart Clinics.

Κράτος θανάτου καὶ ζωῆς

imperium Mortis et Vitae

LONDON

JOHN BALE MEDICAL PUBLICATIONS LTD.

85 GREAT TITCHFIELD STREET, W.1.

1939

PREFACE

The main purpose of this volume is to present a brief account of congenital heart disease with special emphasis on those lesions capable of clinical recognition when modern methods are employed. My friends have sometimes suggested that congenital heart disease is largely a matter for the postmortem room, but my own experience has been the reverse. Eight years of work in five clinics devoted to the study of heart disease in children has made it possible to observe more than 350 cases of congenital heart disease, and these observations form the basis of this work.

First Edition 1939

First Steps

The seminal contributions of Gross, Crafoord, Blalock and Taussig soon followed, and the sense of despair that surrounded the hopeless futilities began to dissipate.

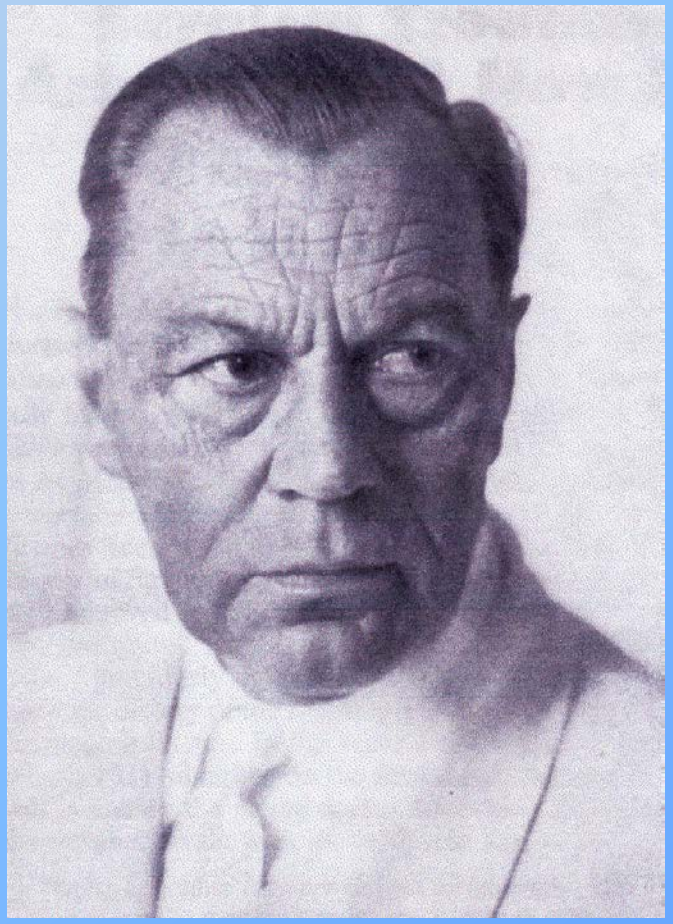
SURGICAL LIGATION OF A PATENT DUCTUS ARTERIOSUS: REPORT OF FIRST SUCCESSFUL CASE

R.E. GROSS

and J. P. HUBBARD

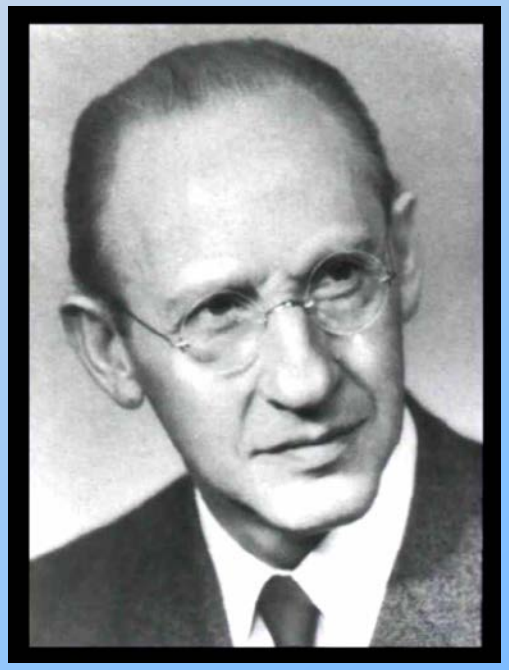
1939

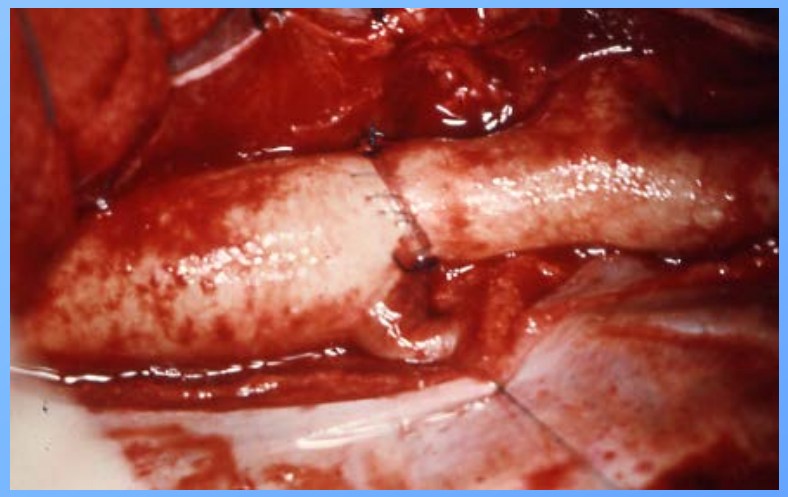
In 1938, E.K. Frey, a surgeon in Dusseldorf, performed the first successful ligation of a patent ductus in a 14 year old boy. Hospital documents were destroyed during the second world war. The case was not reported.



In 1938, Robert Gross and Charles Hufnagel undertook the first experimental studies that anticipated surgical resection of aortic coarctation.

Clarence Crafoord, of Stockholm knew of this experimental work, and while operating on patients with patent ductus arteriosus, “took the risk of placing clamp forceps on the aorta above and below the point of entry of the duct into this artery.” In 1944, Crafoord resected a coarctation and sutured the ends of the aorta together.

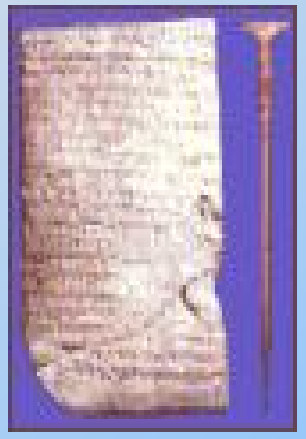




The Pivotal Role of Anesthesia

Anesthesia (Gr "*an*" without + "*aesthesis*" sensation), a term introduced by Oliver Wendell Holmes. Within a span of four years, 1842 to 1846, anesthesia was born and developed in the United States, and ranks among America's greatest contributions to medicine.

The Genesis of Anesthesia was *Genesis*

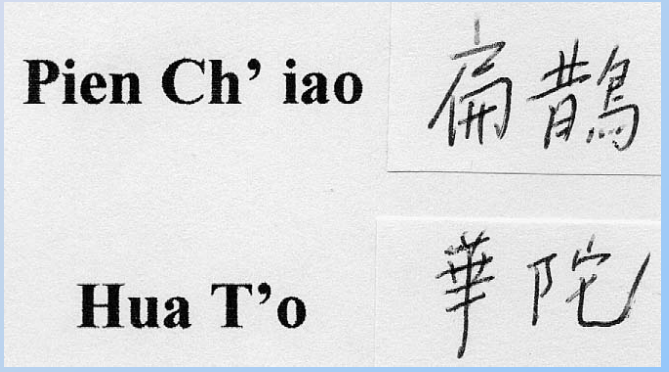


And the Lord God caused a
deep sleep to fall upon Adam, and he slept:
and he took one of his ribs, and closed up
the flesh thereof.

The First Obstetrical Anesthesia

Genesis 2:22 And the rib, which the Lord God had taken from man, made he a woman.

Chinese Medical History

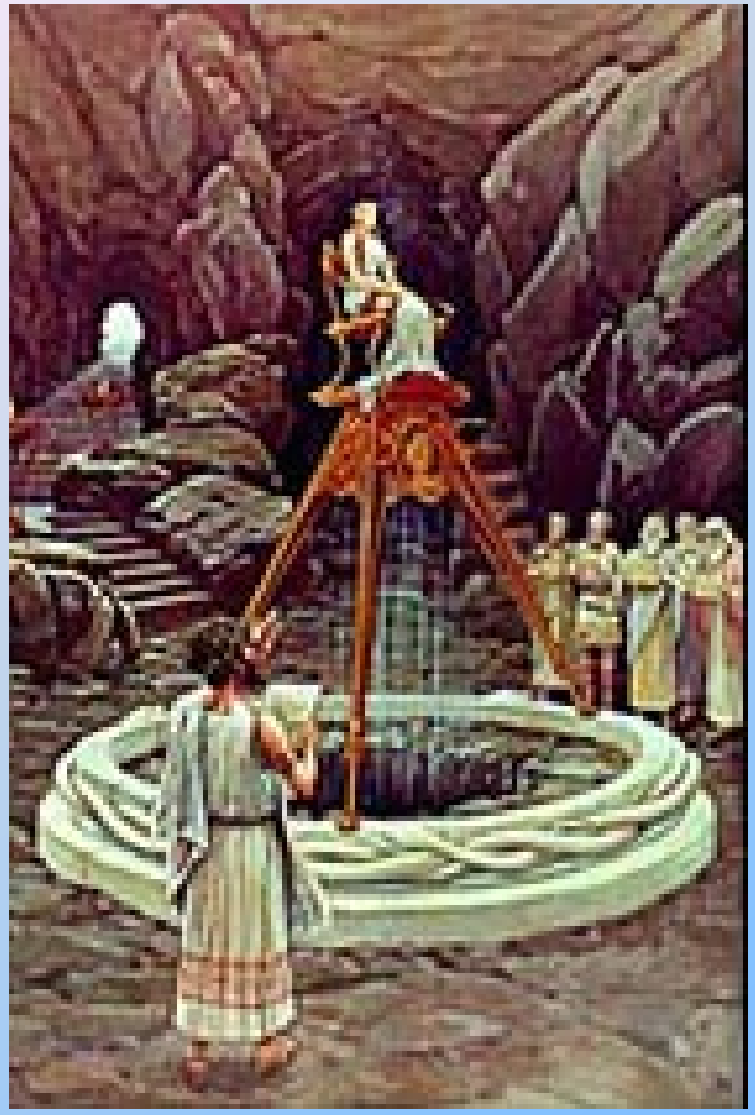


Legend ascribes to Pien Ch'iao in the second century BC the skillful use of anesthesia. The writings of Hua T'o on surgery and anesthesia date from 190 AD.

Temple of Apollo in the Sanctuary of Delphi

Subterranean Anesthesia

The Delphic Oracle



The oracle answered questions while in a trance-like state. Her tripod was directly above a geographic fault from which arose fumes of ethylene which was subsequently used as an anesthetic.

Two Friars

Friar Theodoric of Lucca (1205-98) discovered the use of narcosis in substances that produced sleep during operations for wounds.

Friar Lawrence to Juliet

"Take thou this vial, being then in bed,
And this distilled liquor drink thou off;
When presently through all thy veins shall run
A cold and drowsy humour
And in this borrow'd likeness of shrunk death
Thou shalt continue two and forty hours,
And then awake as from a pleasant sleep."

A Country Doctor

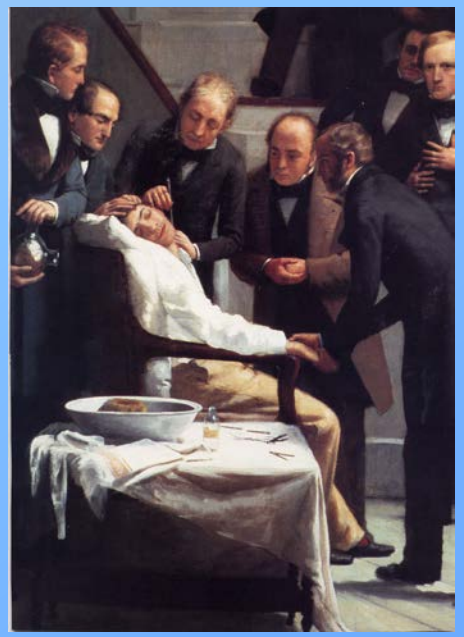
. Ether was first used in surgery on March 30, 1842 by Crawford W Long, a modest country practioner in Jefferson County, Georgia. Seven years elapsed before Long published his observations which received little attention, although no greater boon has ever come to mankind than the power thus granted to induce a temporary but complete insensibility to pain.

In 1842, Crawford Long gave ether to a patient and performed an operation on him. The bill was $2 for the operation and 25 cents for the ether.

Cover design
A Sketch of Medicine
and Pharmacy (1943)

The Ether Dome

William Thomas Green Morton used ether anesthesia at the Massachusetts General Hospital in October 1846. Within a month, the event was published in the Boston Medical and Surgical Journal. Within another month, ether was used in London, and in the succeeding year became widespread throughout Europe.



Anesthetic Device Nineteenth Century.
Science Museum, Oxford

Levy SE, Blalock A. Experimental observations on the effects of connecting by suture the left main pulmonary artery to the systemic circulation. J Thorac Surg 1939.

In 1944, Alfred Blalock sutured the end of a subclavian artery to the side of a pulmonary artery in a patient with Fallot's tetralogy, establishing the now legendary Blalock-Taussig anastomosis, proving that a deeply cyanosed child could tolerate operation and improve substantially.

The Blalock/Taussig Shunt

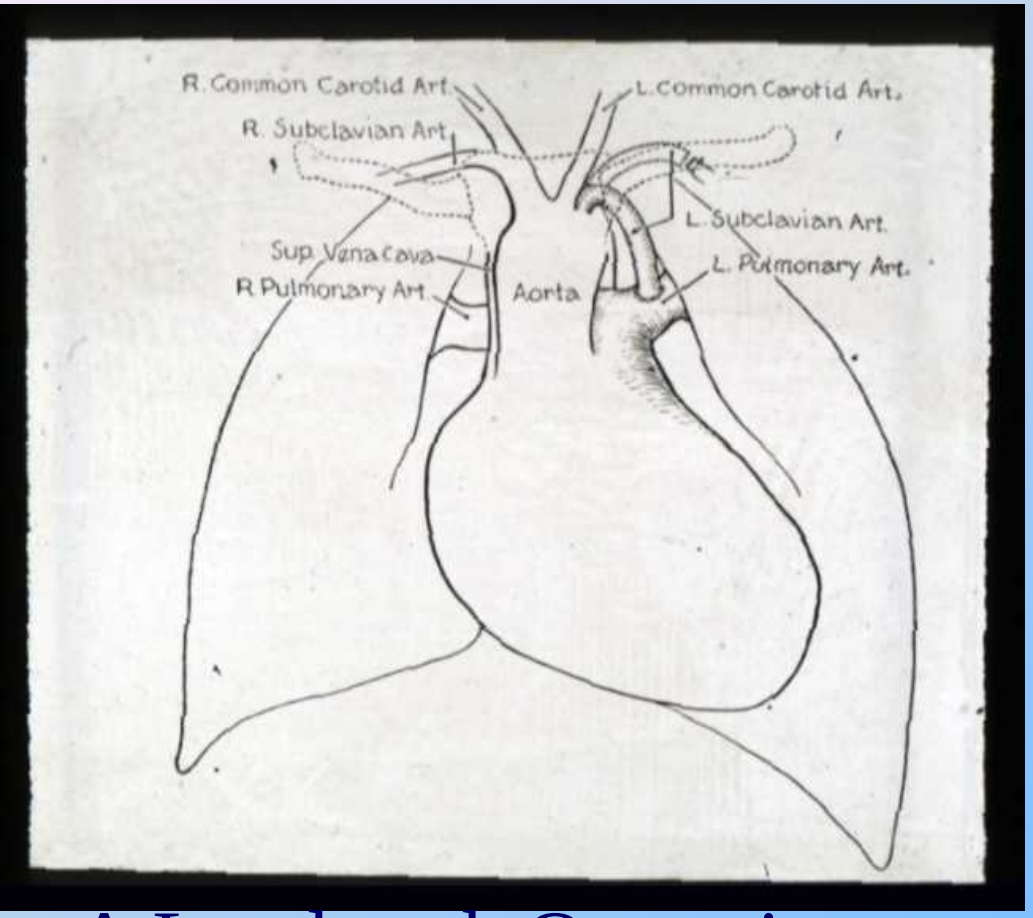
The anesthesiologist for the seminal operation was Dr Merel Harmel, a junior faculty member at Johns Hopkins, who used open drip ether. Dr Austin Lamont, Chief of Anesthesiology, had been asked by Dr Blalock to give the anesthesia, but Lamont refused, commenting, “I will not put that child to death.”

Merel Harmel

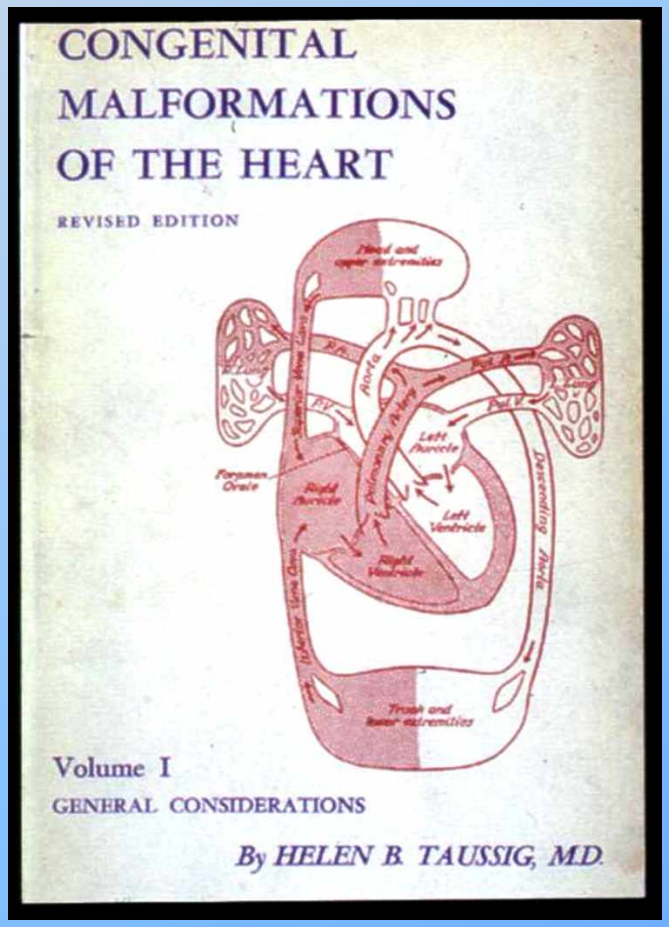
Eileen Saxon

Alfred Blalock

Helen Taussig

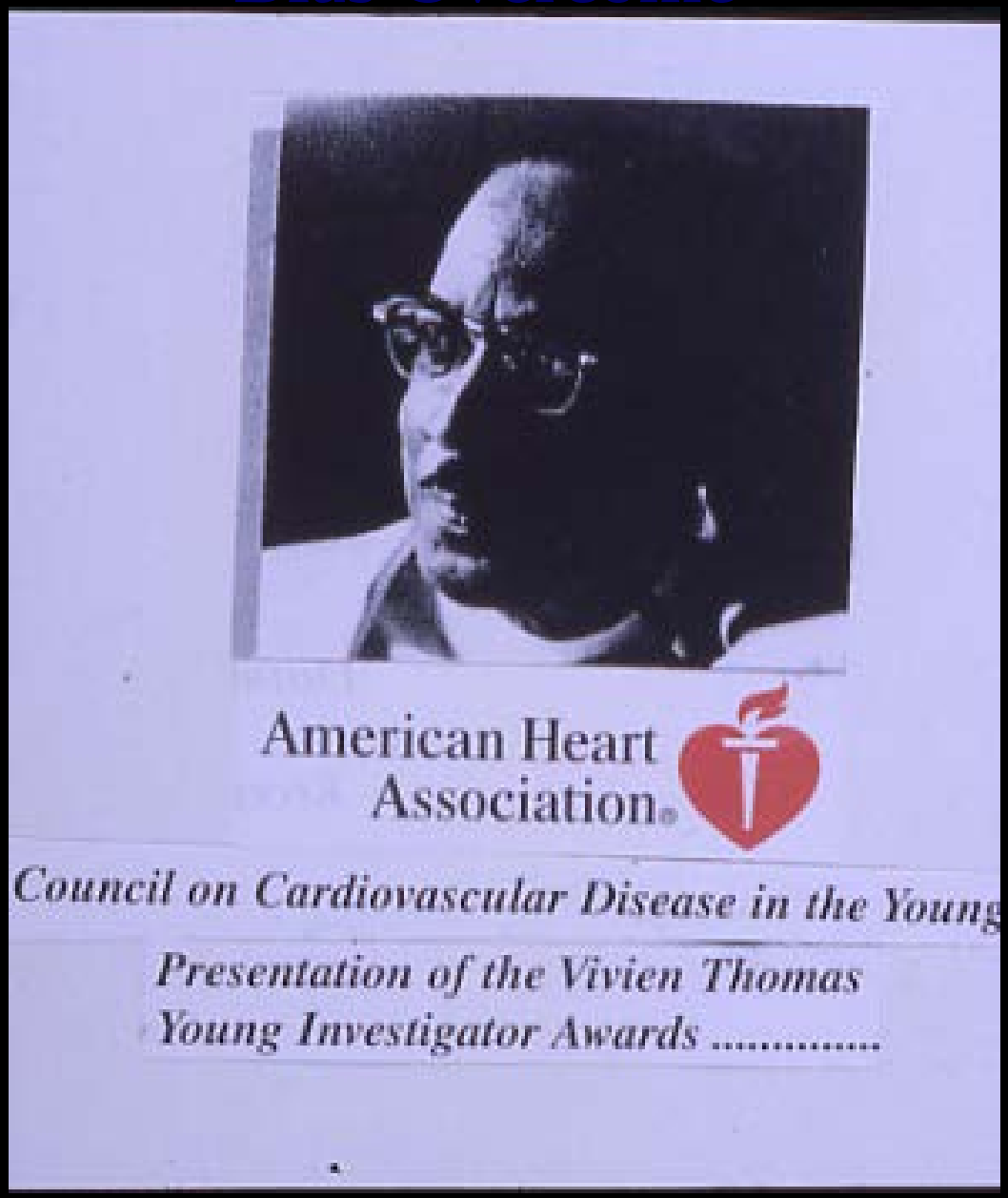


A Landmark Operation
1944



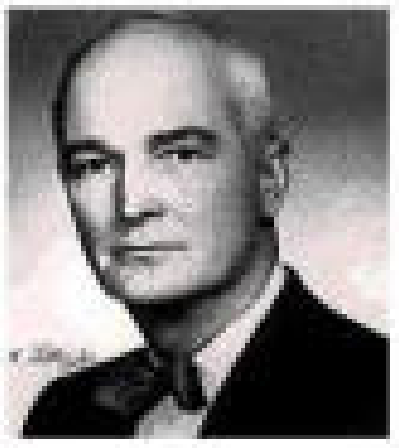
A Landmark Book
1947

Bias Overcome



The Gibbon Pump

"…the patient's life might be saved if some of the blue blood in her veins could be continuously withdrawn into an extracorporeal blood circuit, exposed to an atmosphere of oxygen, and then returned to the patient by way of a systemic artery. Cardiorespiratory function might be temporarily performed by the blood circuit while the massive pulmonary embolus was surgically removed."



John H. Gibbon

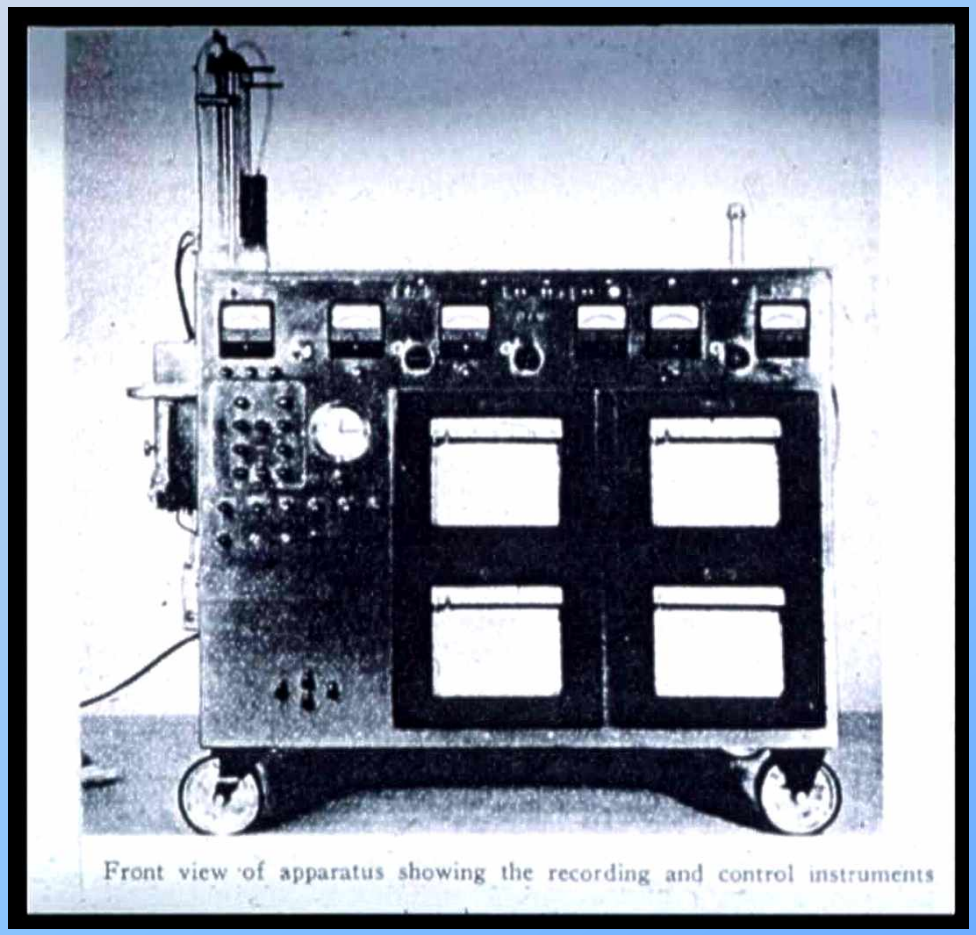
Surgical Fellow

Massachusetts General Hospital, 1931

Minnesota Med. 37:171,1954

APPLICATION OF A MECHANICAL HEART AND LUNG APPARATUS TO CARDIAC SURGERY

JOHN H. GIBBON, Jr., M.D.
Philadelphia, Pennsylvania



Front view of apparatus showing the recording and control instruments

The first successful intracardiac operation (1953) employed the Gibbon heart/lung bypass system for closure of an atrial septal defect in an 18-year-old woman.

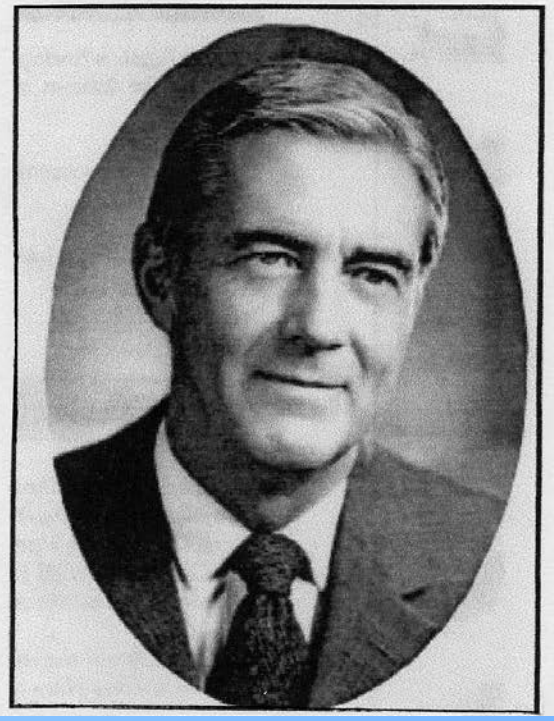
HEART AND LUNG APPARATUS
Contemporary

Wilfred Gordon Bigelow

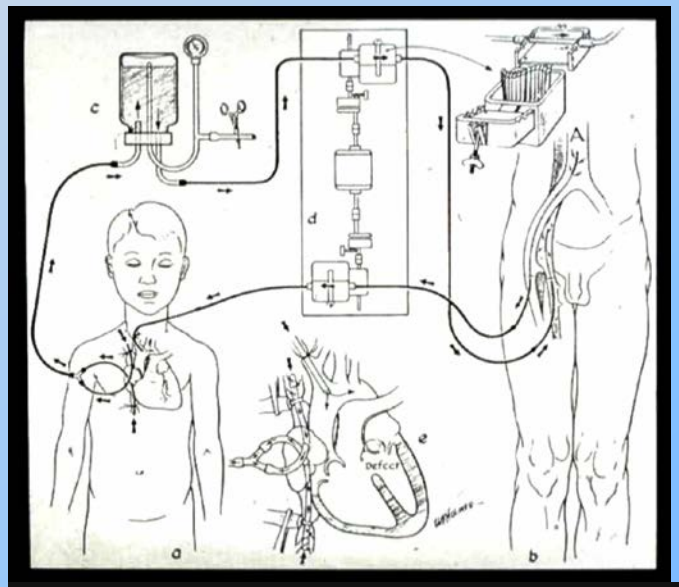
1941 — First interest in hypothermia research

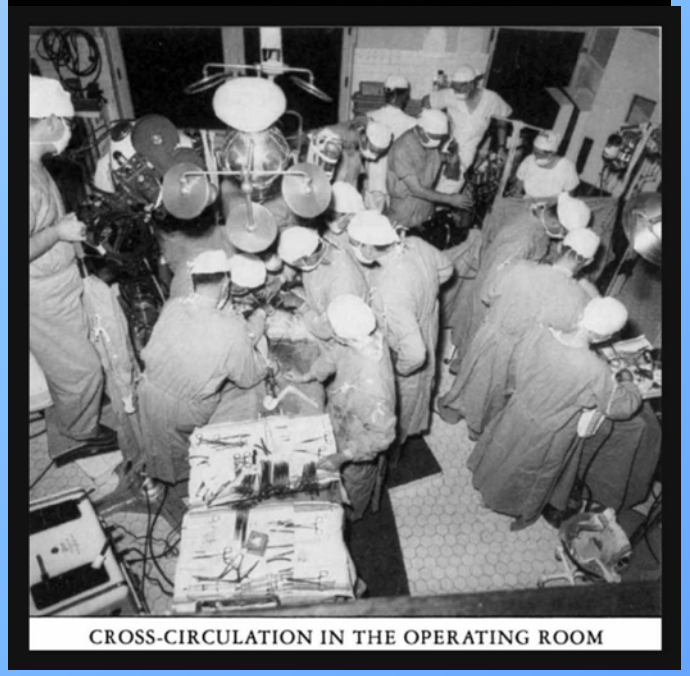
1951 — The pacemaker which evolved unexpectedly out of his hypothermia research

1953 — First successful human application of hypothermia for open heart surgery

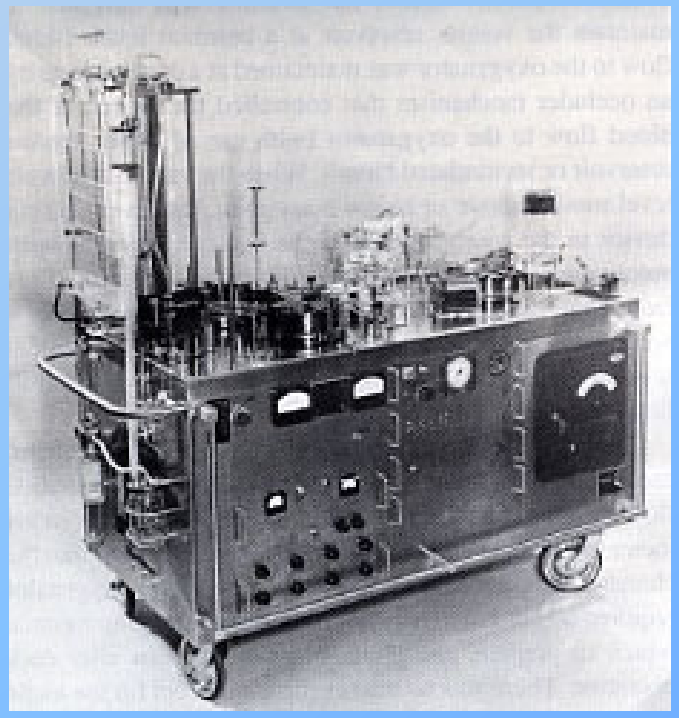


1955

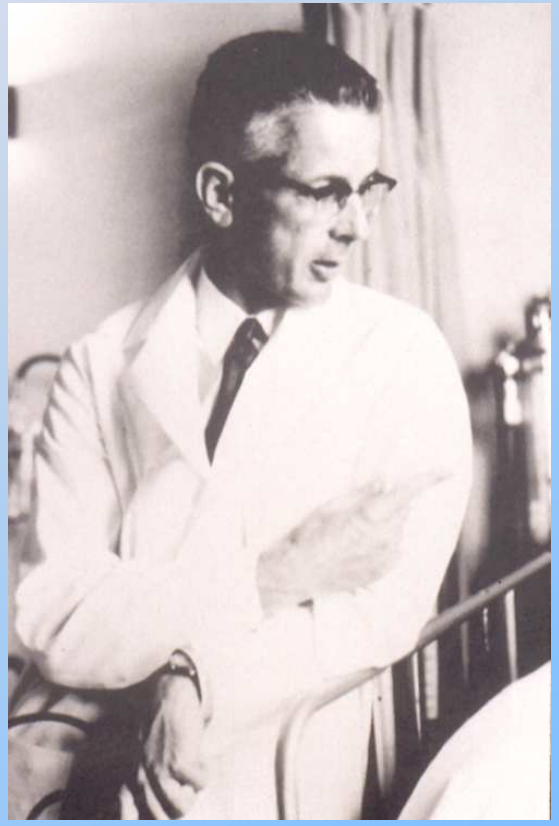



CROSS-CIRCULATION IN THE OPERATING ROOM

John W. Kirklin



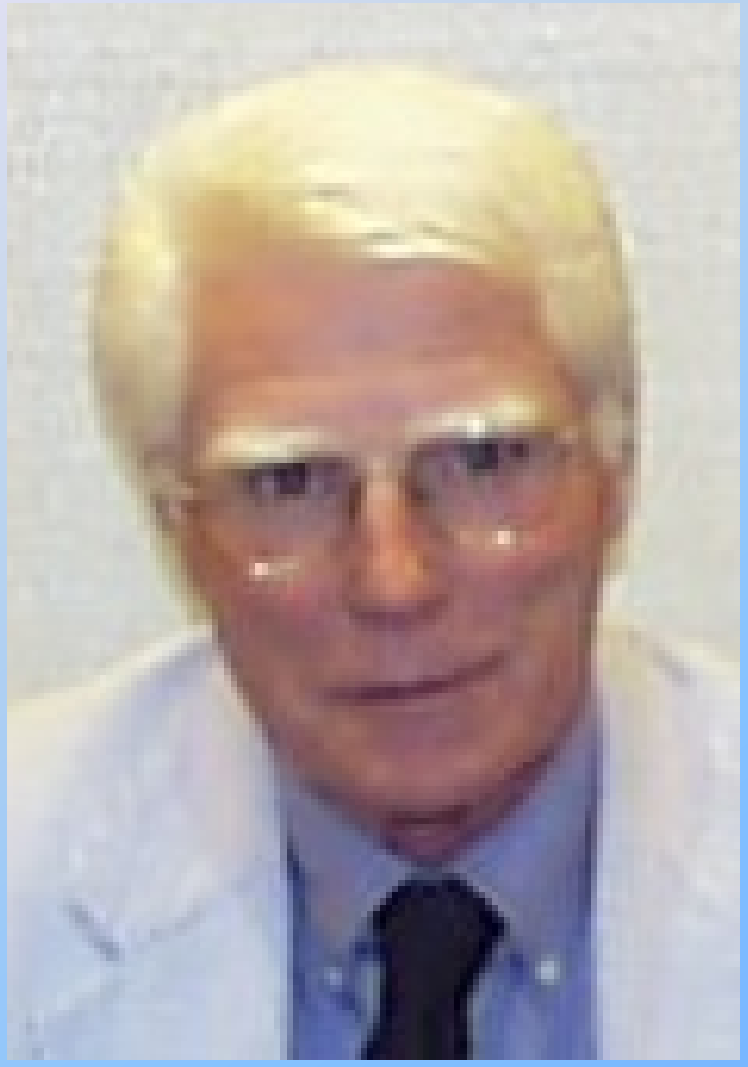
Heart-lung machine used
at the Mayo Clinic in March 1955



INTRACARDIAC SURGERY WITH THE AID OF A MECHANICAL PUMP-OXYGENATOR SYSTEM (GIBBON TYPE): REPORT OF EIGHT CASES*

John W. Kirklin, M.D., Section of Surgery

Proc. Staff Meetings Mayo Clinic **30**:201–206 (1955)

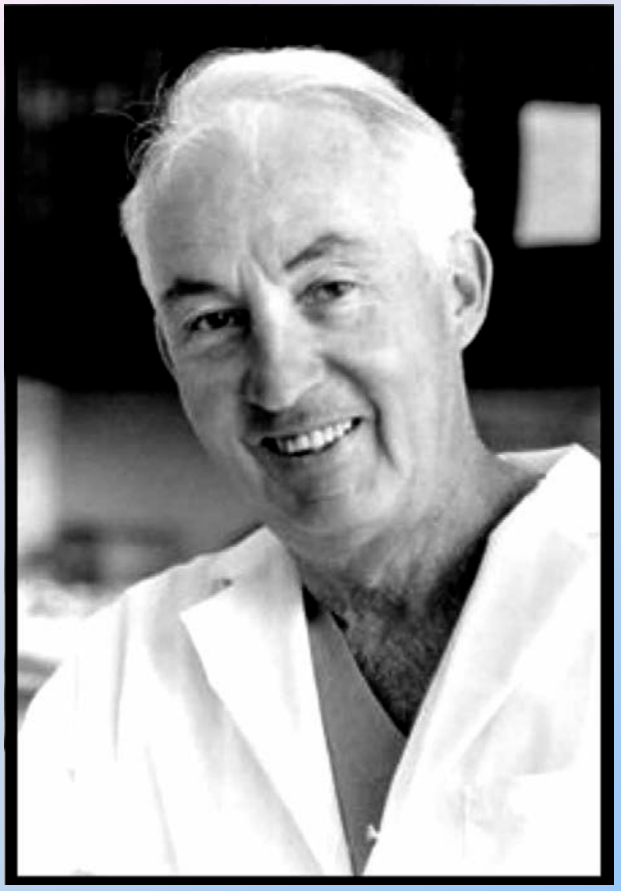


Aldo Casteneda, MD,PhD
Cardiac Surgery in the Neonate and Infant 1994

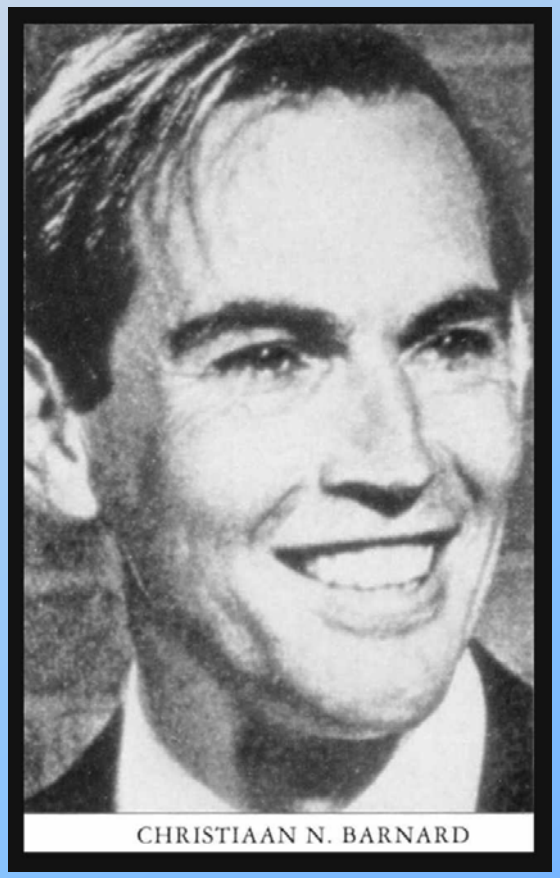
The First Cardiac Transplantation

Pien Ch’iao practiced in China in the second century BC. Legend ascribes to him such skillful use of anesthesia that he was able to operate painlessly and even exchange the hearts of two patients.

Nobel laureate Alexis Carrel demonstrated in 1905 that a canine heart could be heterotopically transplanted and resume function in the new host.

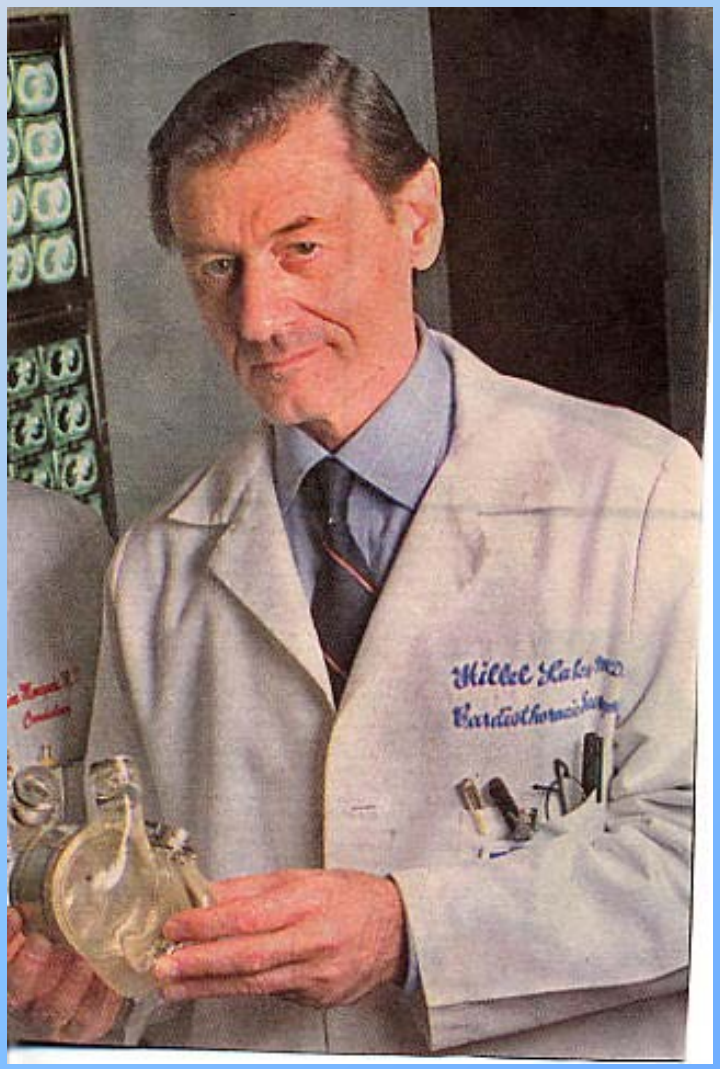


1960 — Lower and Shumway reported the first successful orthotopic cardiac transplantation.



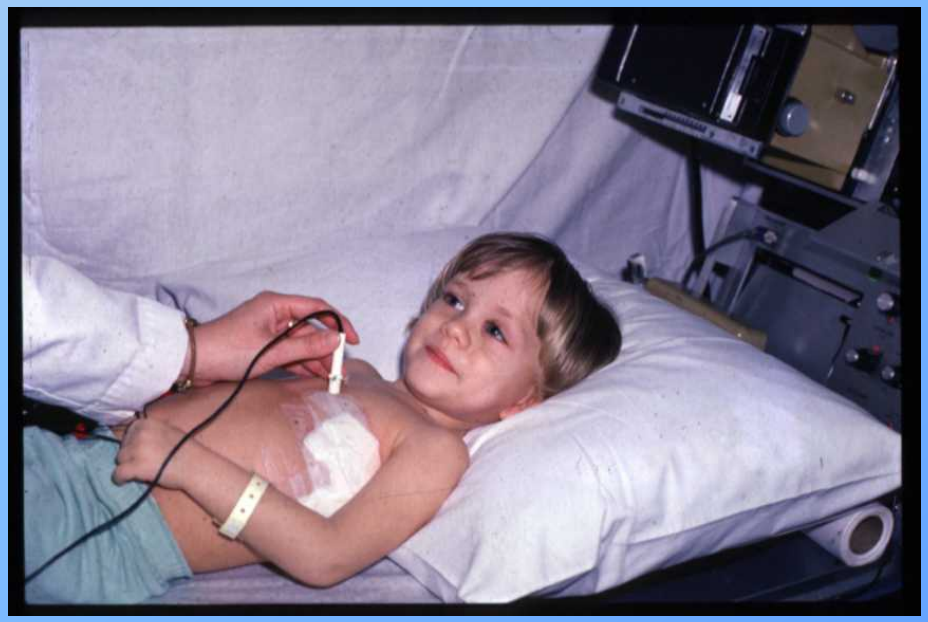
1966 — Christian Barnard performed the first human cardiac transplantation.

The Mechanical Heart
Hillel Laks

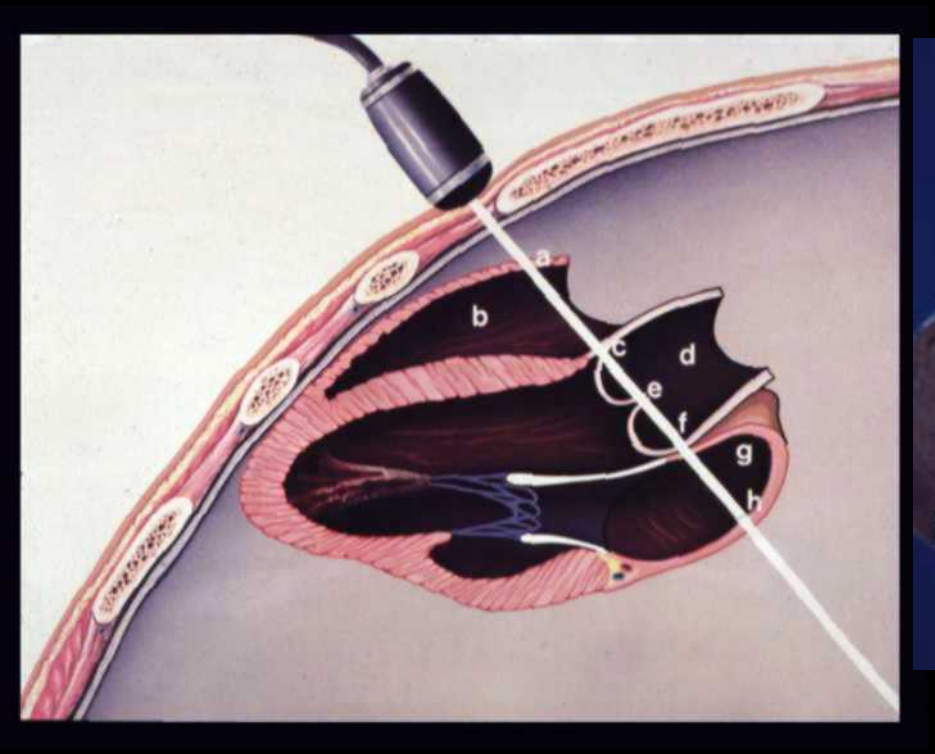


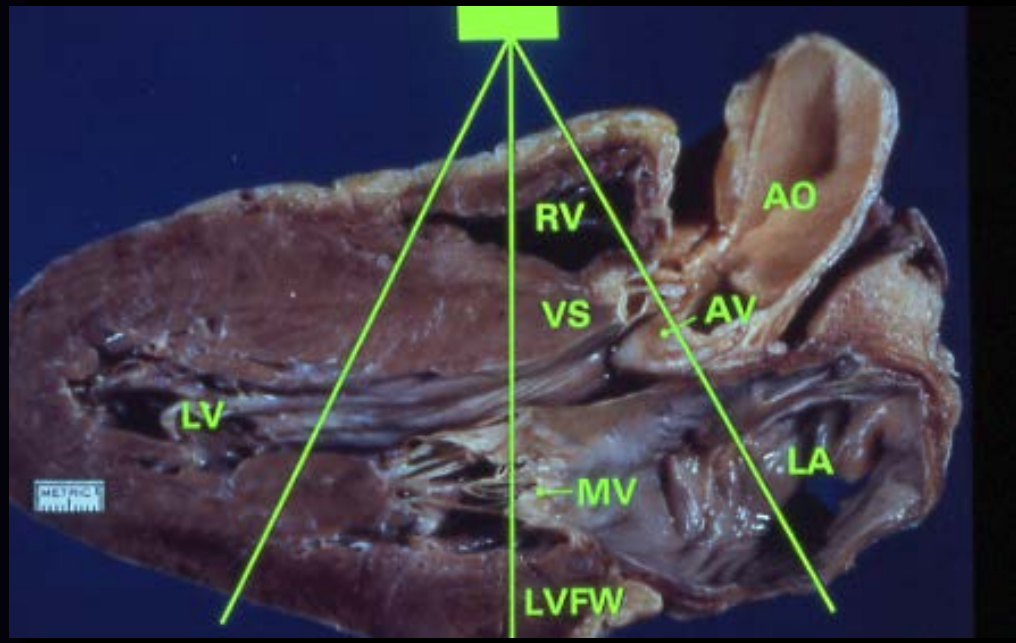
The Diagnostic Pathway to the Promised Land

Echocardiography began with the discovery of piezoelectricity in 1880 by Pierre and Jacques Curie

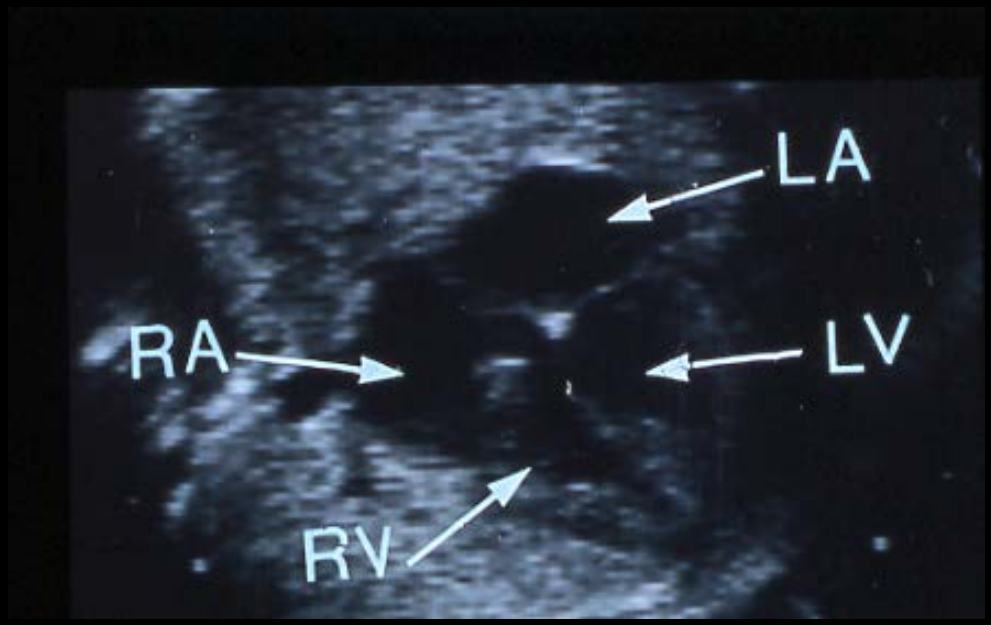


Transthoracic

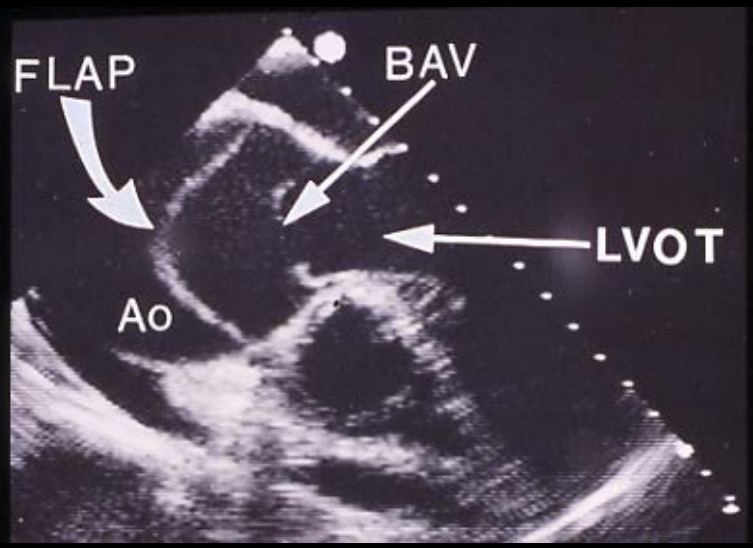




Fetal

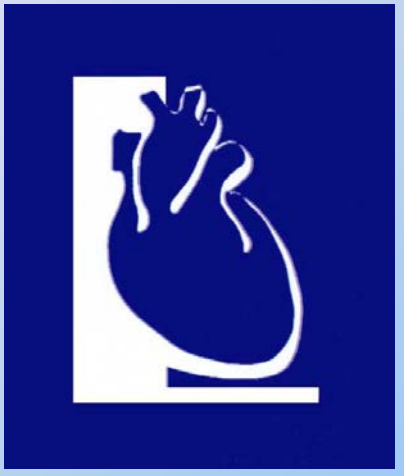


Transesophageal



Gadolinium-Enhanced Magnetic Resonance Image

Hemitruncus



Cardiac Catherization

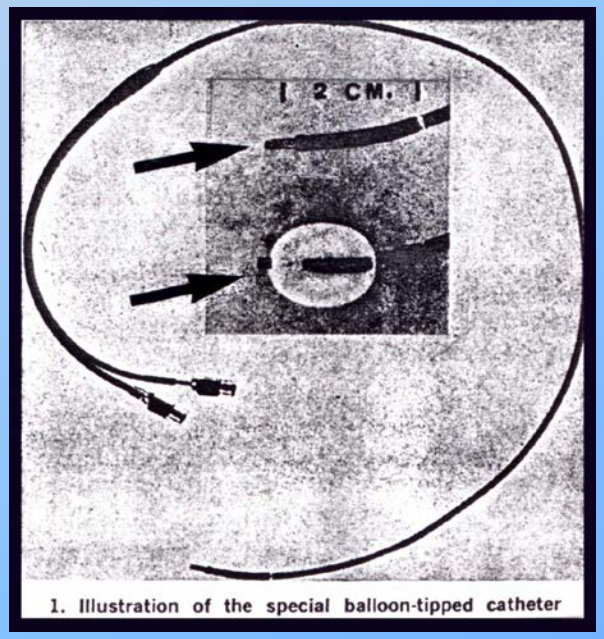
A Therapeutic Intervention

Am. Med. Assoc. J. 1966

Creation of an Atrial Septal Defect Without Thoracotomy

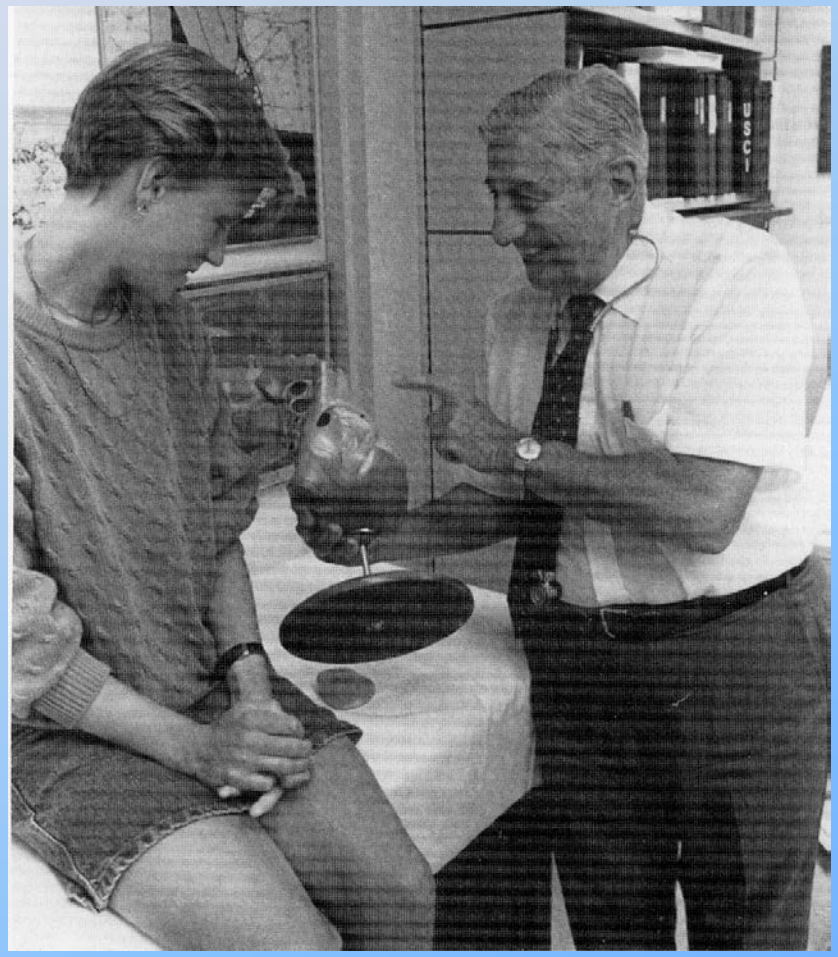
A Palliative Approach to Complete Transposition of the Great Arteries

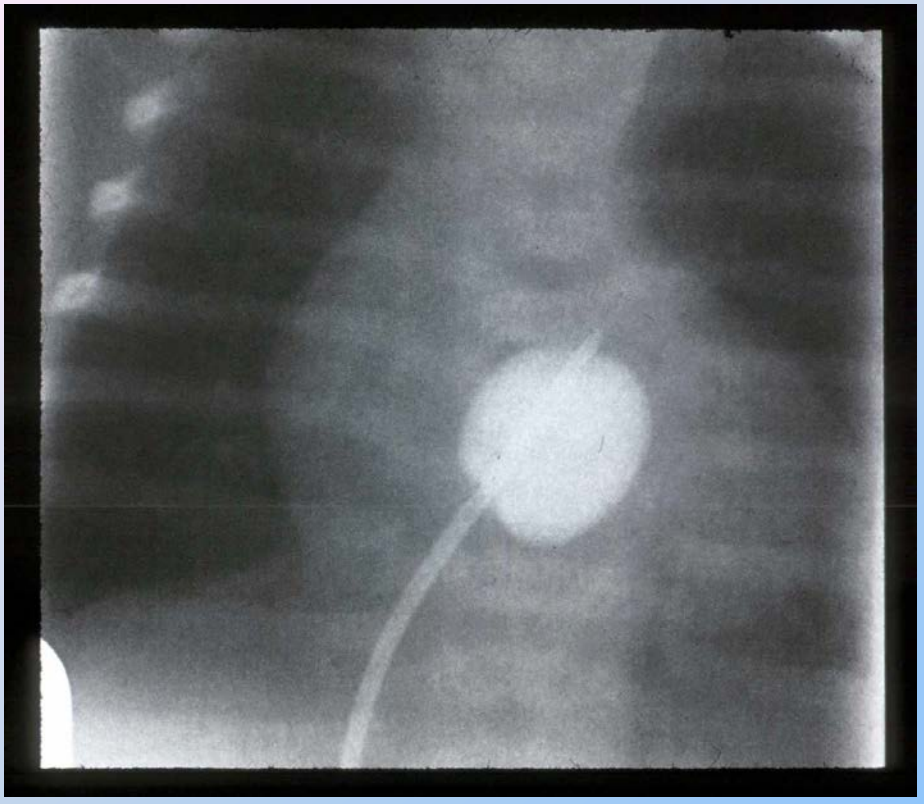
William J. Rashkind, MD, and William W. Miller, MD

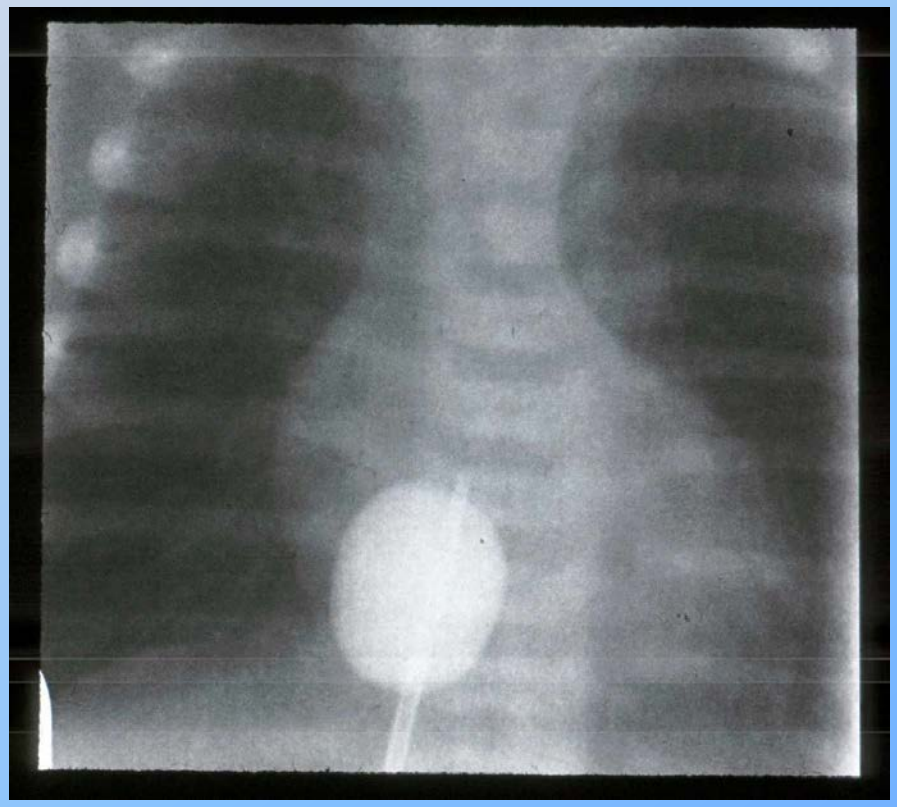


1. Illustration of the special balloon-tipped catheter

William J. Rashkind



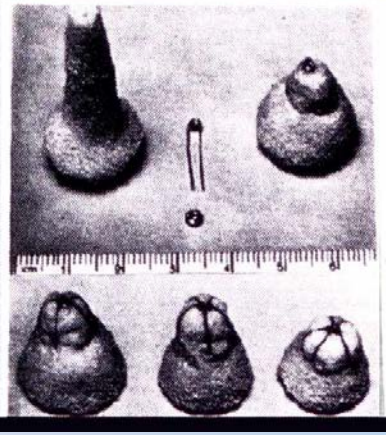




Success depends on the jerk at the end of the catheter

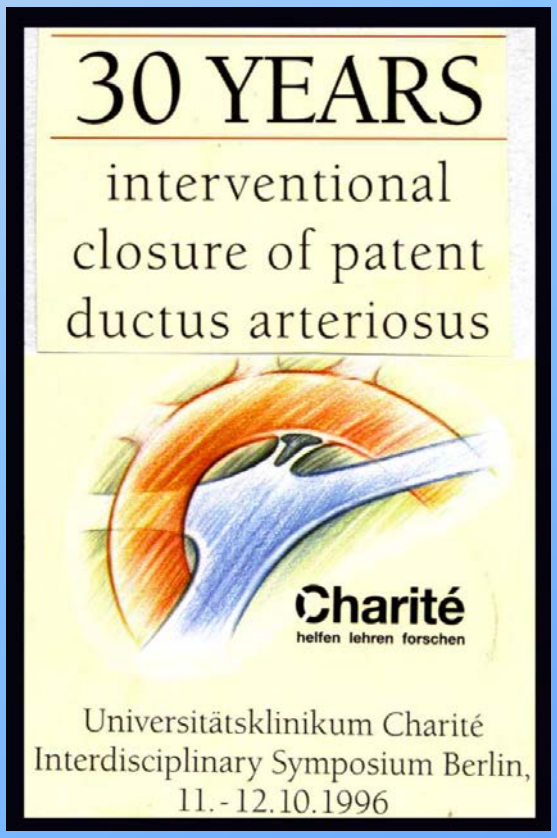
WJR

The first publication about the interventional closure of patent ductus arteriosus in 1967.



W. Porstmann; L. Wierny; H. Warnke
Der Verschluß des Ductus arteriosus persistens ohne Thoraktomie (vorläufige Mitteilung) Thoraxchirurgie Vaskuläre Chirurgie 15: 199-203 Stuttgart (1967)

Portsmann (2nd left), Forsmann (2nd right)



The Future of Congenital Heart Disease

Despite remarkably precise anatomic and physiologic diagnoses and astonishing surgical feats, cures in the literal sense are few. We are therefore obliged look beyond the present and anticipate responsibility for the long-term care of new generations of patients with the sequelae and residua of postoperative congenital heart disease.

Long Term Follow-Up.
By Whom ?

The relative geographic mobility of populations in the United States makes it unlikely that patients with congenital heart disease will remain under the long term care of their pediatric cardiologists.

Current Resources in the US

Subspecialty Board Certification

- Pediatric Cardiology ---------- 1,200
- Cardiovascular Medicine -----20,000

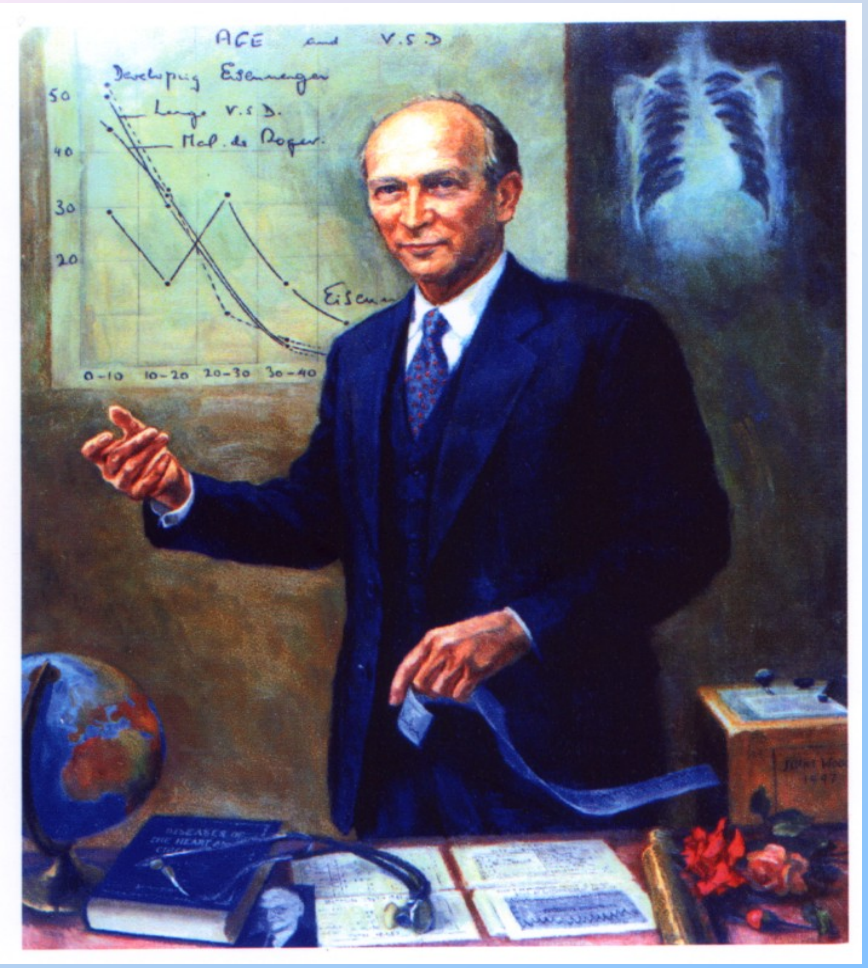
Specialized Tertiary Care for Adults With CHD

Specialized facilities for the comprehensive care of adults with congenital heart disease do not compete with practicing physicians or community hospitals, but instead offer services difficult if not impossible to duplicate.

Personal Reminiscences

Turning the efforts of many years into an hourglass

The 1950's



Portrait of Paul Wood
by his daughter, Juliet

DISEASES OF
THE HEART AND
CIRCULATION

by

PAUL WOOD, O.B.E.

M.D. (Melbourne), F.R.C.P. (London)

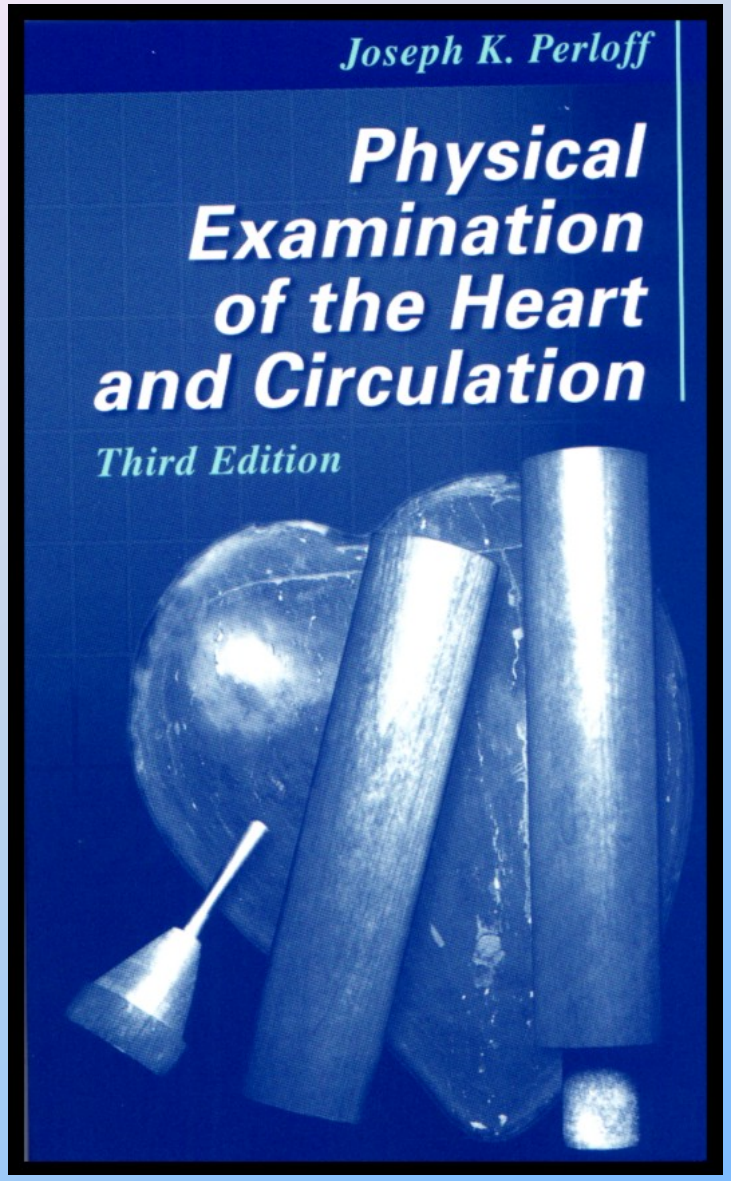
Director, Institute of Cardiology, London
Physician, National Heart Hospital
Physician in charge of the Cardiac Department, Brompton Hospital

CHAPTER II. PHYSICAL SIGNS

CHAPTER VIII. CONGENITAL HEART DISEASE

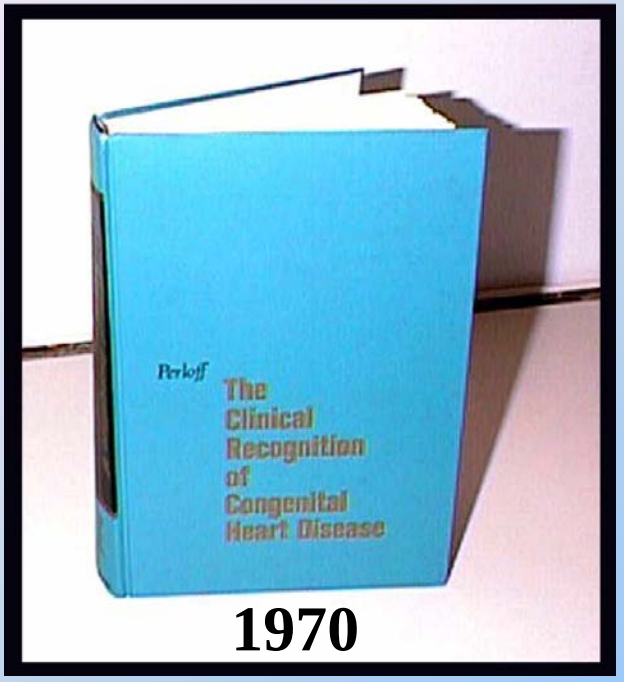
London 1954


THE KING'S HEAD
WINES
SPIRITS
CHARRINGTON'S
WHEATLEY
STREET W.1



To the memory of
PAUL HAMILTON WOOD
from whom I learned
the physical examination
of the heart and
circulation.

PREFACE



1970

This book does not confine itself to congenital cardiac disease in either children or adults, but includes all ages in order to give a complete, unified and uninterrupted impression of each malformation.

Congenital Heart Disease
A Continuum

Neonates

Infants

Children

Adolescents

Adults

The Early 1970's

45th Scientific Sessions, American Heart Association, Dallas, Texas, November, 1972

Pediatric Congenital Cardiac Becomes a Postoperative Adult

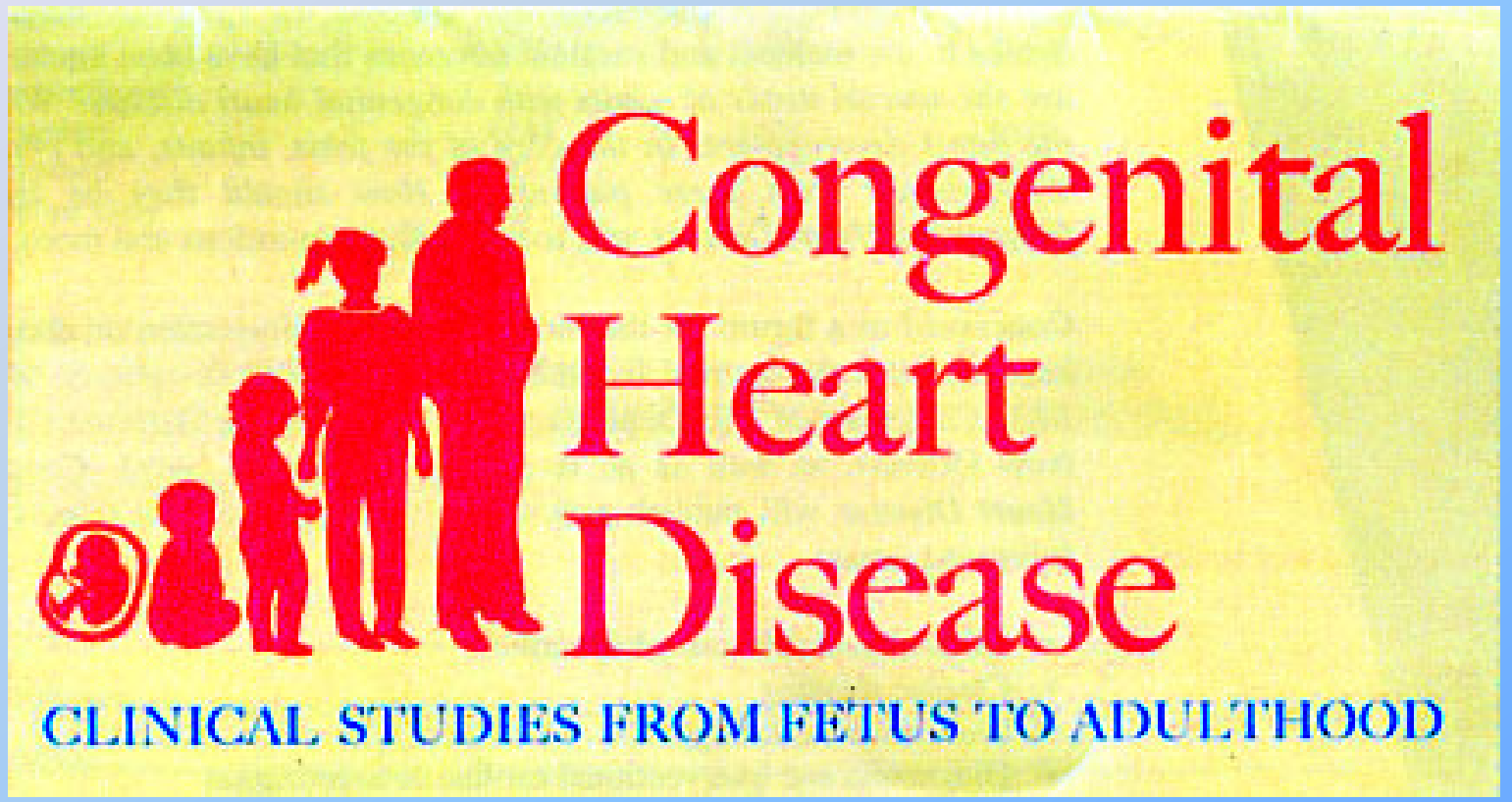
The Changing Population of Congenital Heart Disease

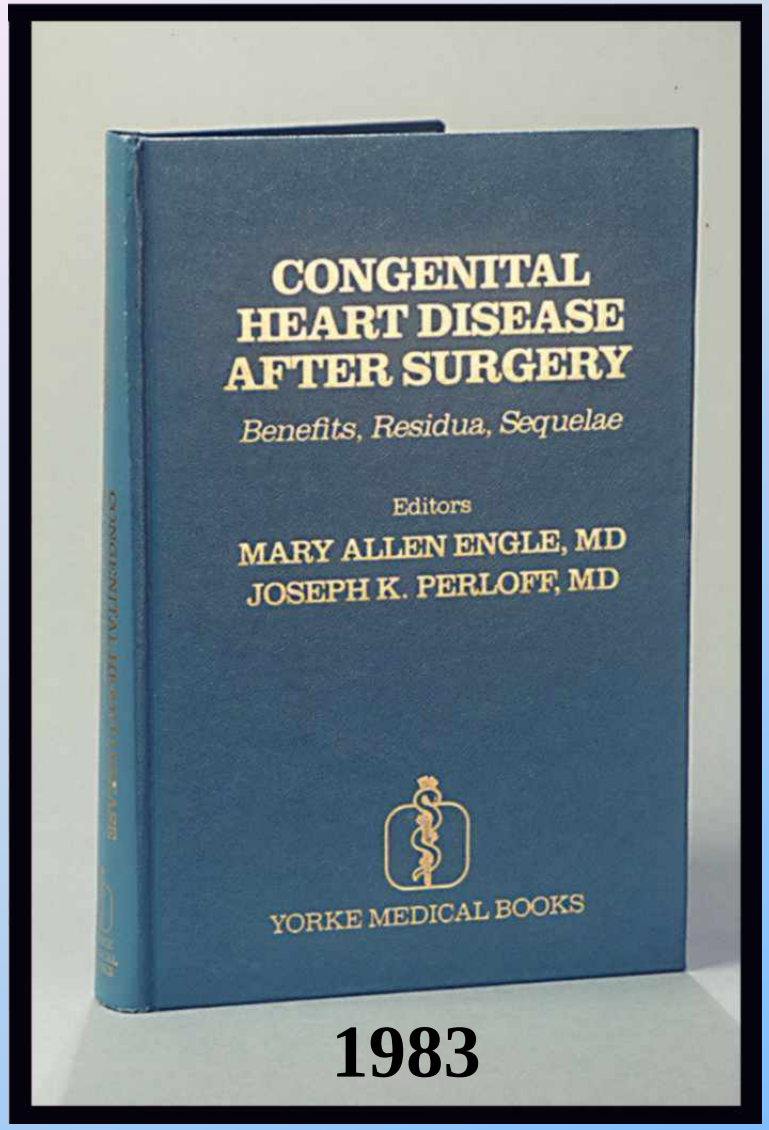
Joseph K. Perloff

Circulation, March 1973

2006

The Changing Population Has Changed





With admiration and affection we dedicate this book to Dr. Helen Brooke Taussig.

Mary Allen Engle

Joseph K. Perloff

World Survey of the Common Cardiac Malformations: Developmental Error or Genetic Variant?

Helen B. Taussig, MD

Ahmanson/UCLA Adult Congenital Heart Disease Center

Congenital Heart Disease in Adults
The Future of Children

The Infant

The Postoperative Adult

Adult Congenital Heart Disease in the United States

Simple, moderately complex, complex:

900,000 to 1,000,000

20,000 open operations per year

Annual increase approximately 5% per year

There are more adults with CHD in the US than there are infants and children.

Worldwide

1.5 million new cases per year

Approximately 85% of afflicted infants in developed countries reach adulthood

BETHESDA CONFERENCES

22nd Bethesda Conference, October 1990

Congenital Heart Disease After Childhood:

An Expanding Patient Population

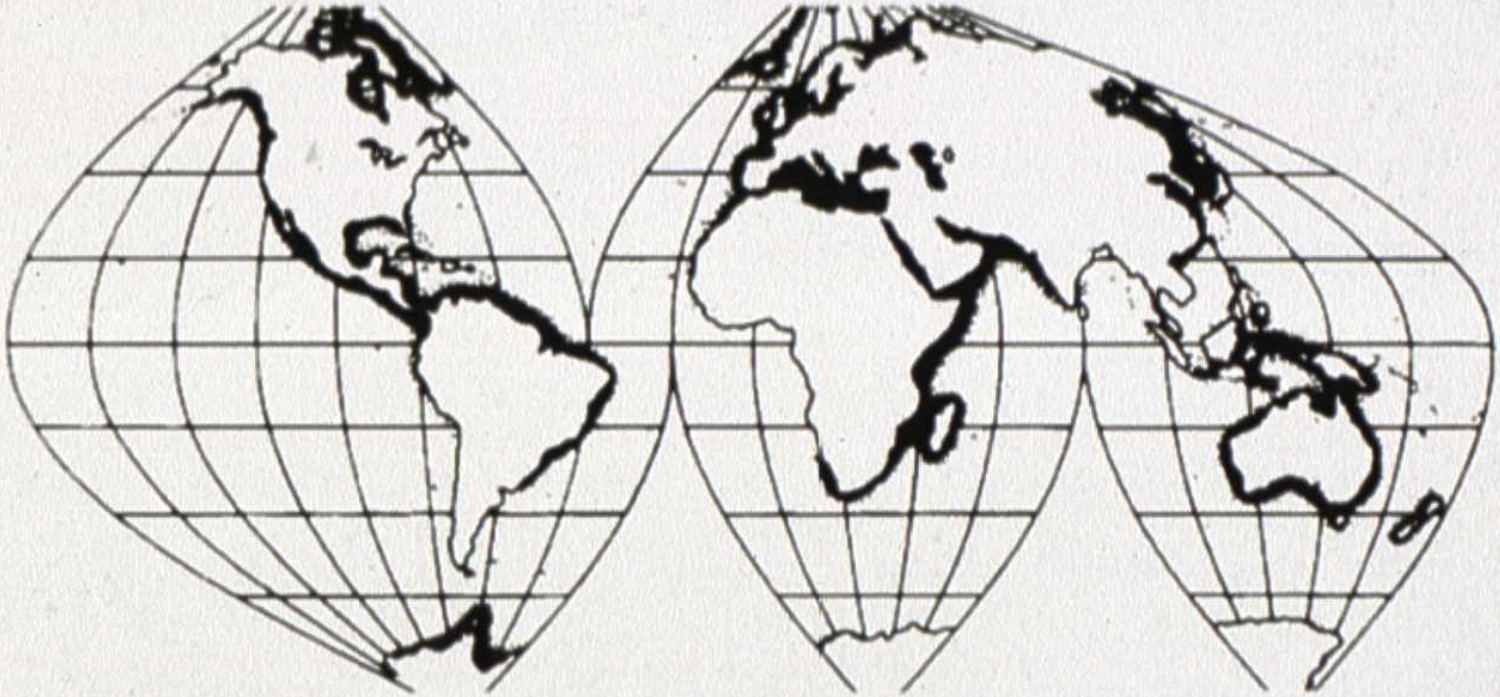
JOSEPH K. PERLOFF, Conference Chair

32nd Bethesda Conference, October 2000

Care of the Adult with Congenital Heart Disease

Gary Webb and Roberta G. Williams, Conference Chairs

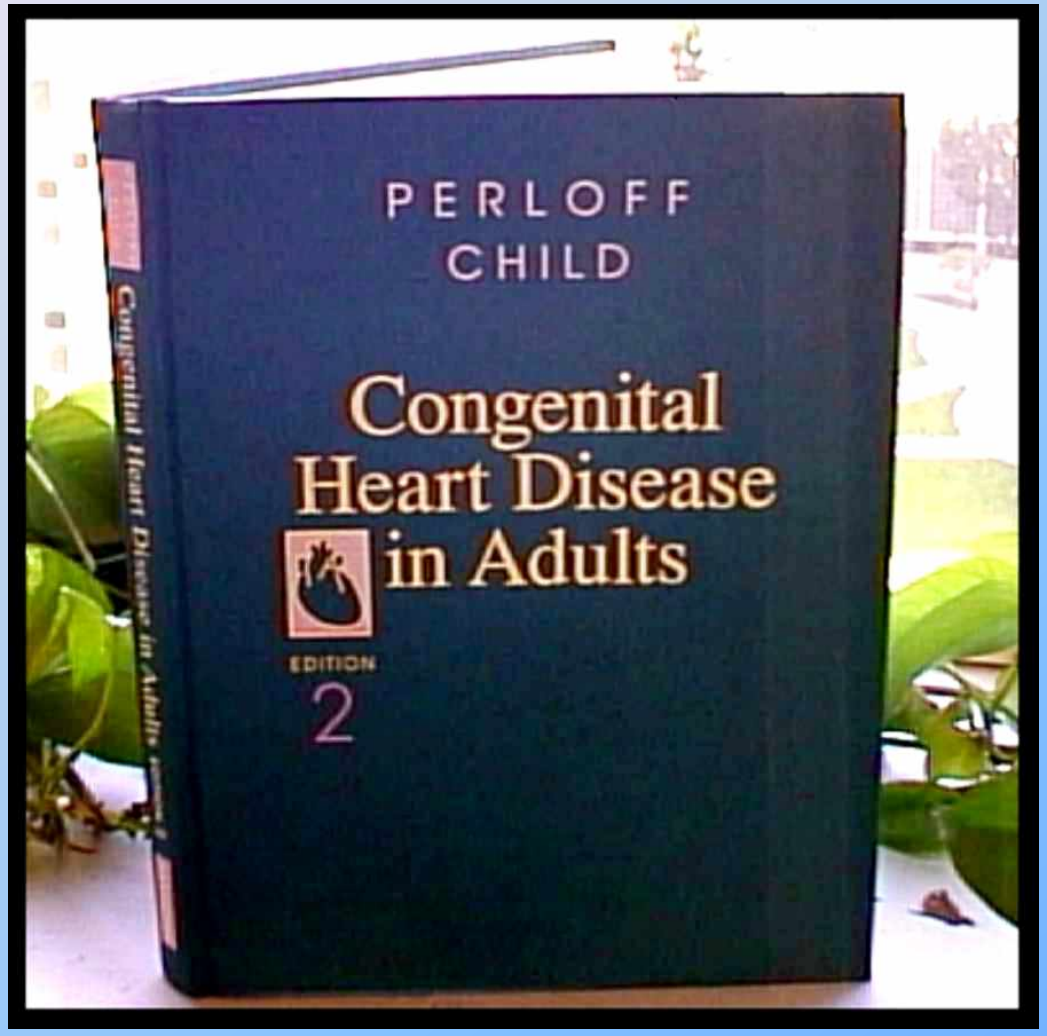
International Society for



Adult Congenital Cardiac Disease

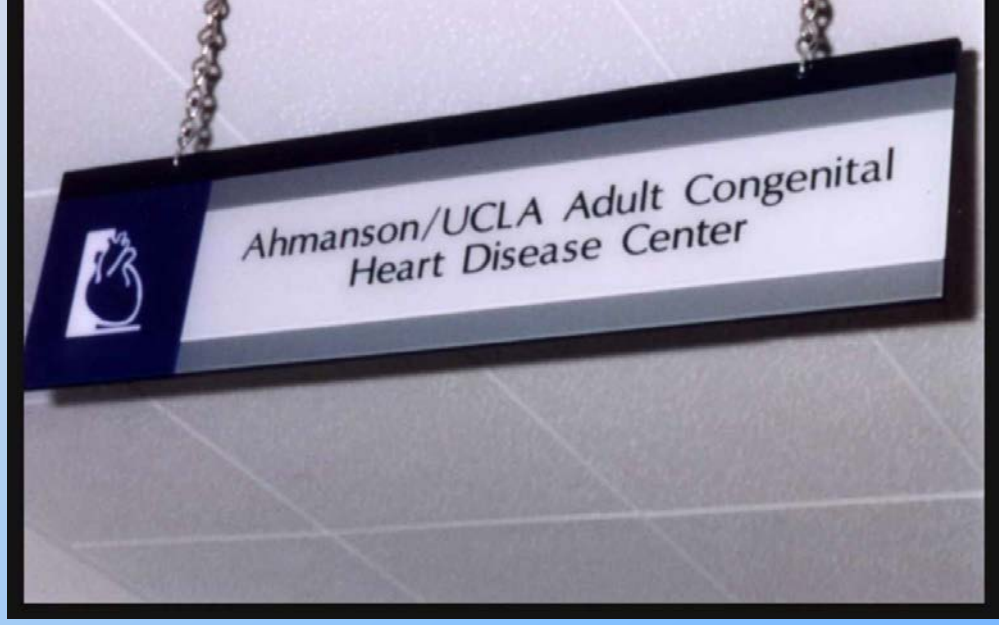
A Patient Advocacy Group

Resurrection


PERLOFF
CHILD
Congenital
Heart Disease
in Adults
EDITION
2
Congenital Heart Disease in Adults

The New Age

London 1954
The National Heart Hospital



Los Angeles 2006
The UCLA Hospital

Ahmanson/UCLA Adult Congenital Heart Disease Center

EPILOGUE

Thus far, with rough and all-unable pen,
Our bending author hath pursued the story.

Pediatric cardiology evolved because advances in diagnostic methods and surgical management of CHD created an air of optimism in infants regarded as hopeless futilities. The early years of this fledgling specialty were not easy because entrenched biases weighed heavily against innovation.

EPILOGUE

Nobody Knows the Troubles I've Seen
Nobody Knows My Sorrow

C*ongenital heart disease in adults* evolved because the stunning success of pediatric cardiologists and pediatric cardiac surgeons profoundly changed survival patterns. The early years of this fledgling specialty were not easy because entrenched biases weighed heavily against innovation.

When those of you who are destined to lead look ahead, you will often confront regressive opposition. And when you look back, there may be no one following. But vigor can be derived from opposition. And remember:

This is not the end. It is not even the beginning of the end. But it is perhaps the end of the beginning.