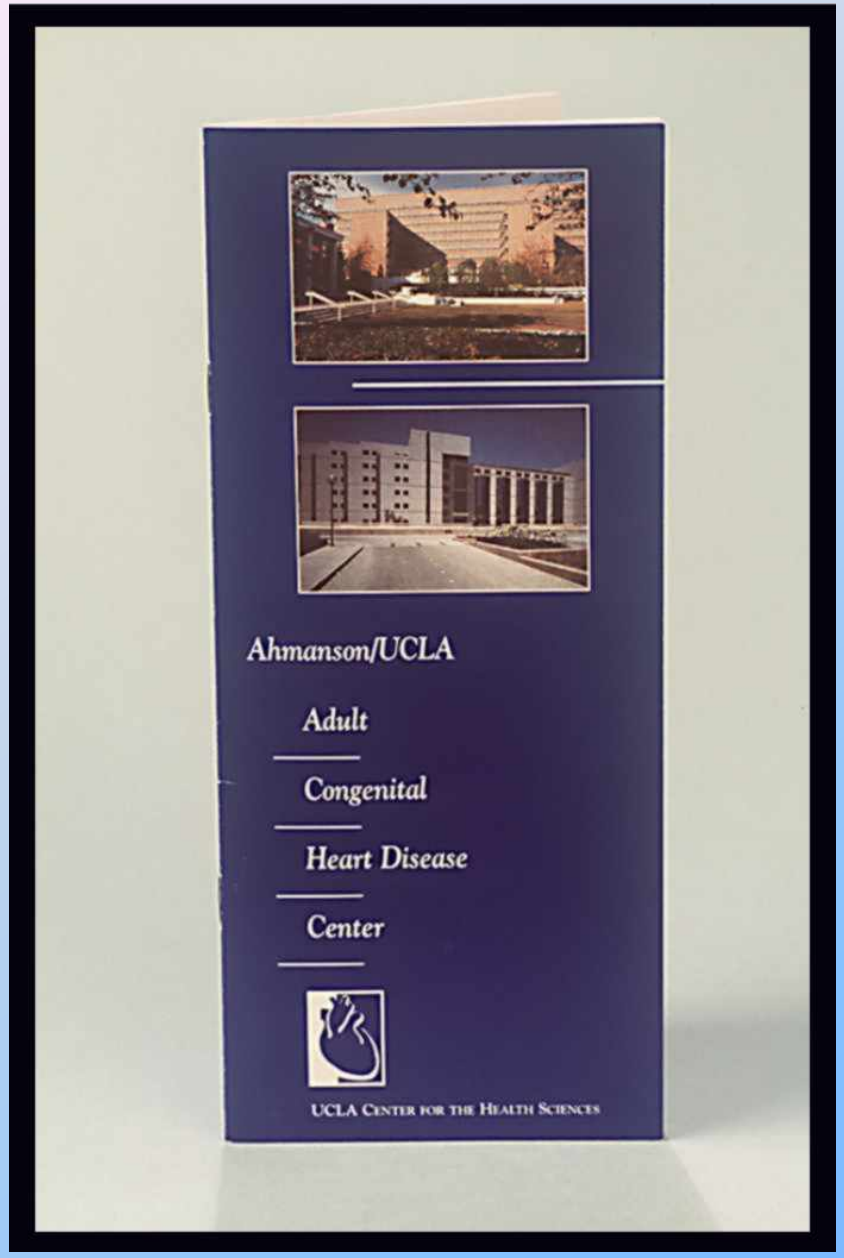
St Luke'sInternational Hospital, Tokyo

Second Adult Congenital Heart Disease Conference 2012



Adult Congenital Heart Disease

Historical Perspectives and Personal Reminiscences

Joseph K. Perloff

Four Decades Ago

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

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

The First Childrens' Hospital

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 the treatment of the young. Until the turn of the 20th century, such 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

1079 pages. The Section on Diseases of the Circulatory System contained a 5 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.

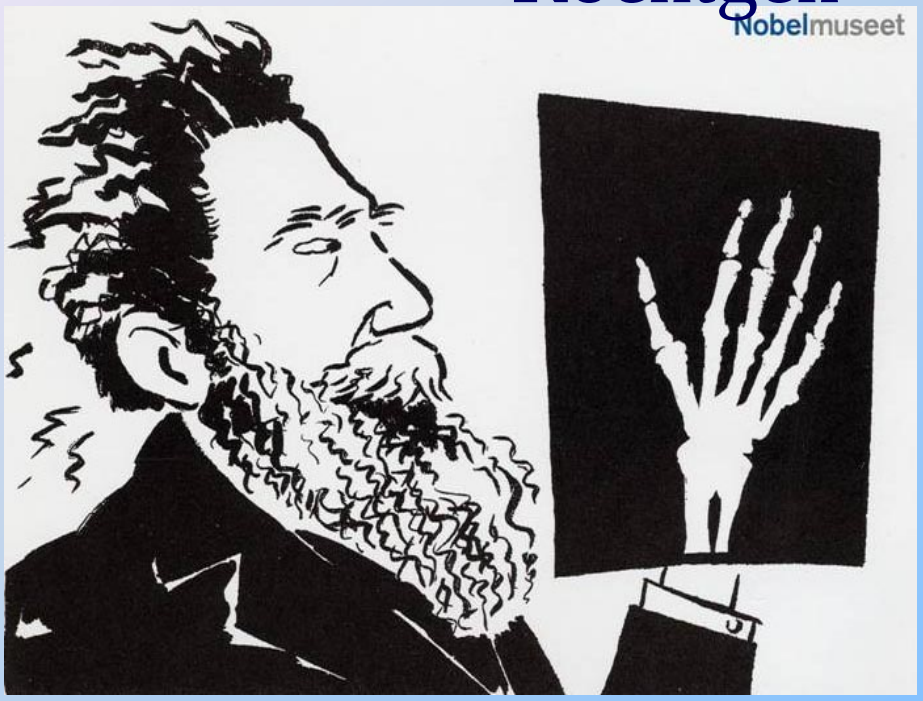
Holt's THE DISEASES OF INFANCY AND CHILDHOOD

1897

1117 pages. The Section on Diseases of the Circulatory System contained a 7 page chapter devoted to *Congenital Anomalies of the Heart.*

1895

Roentgen



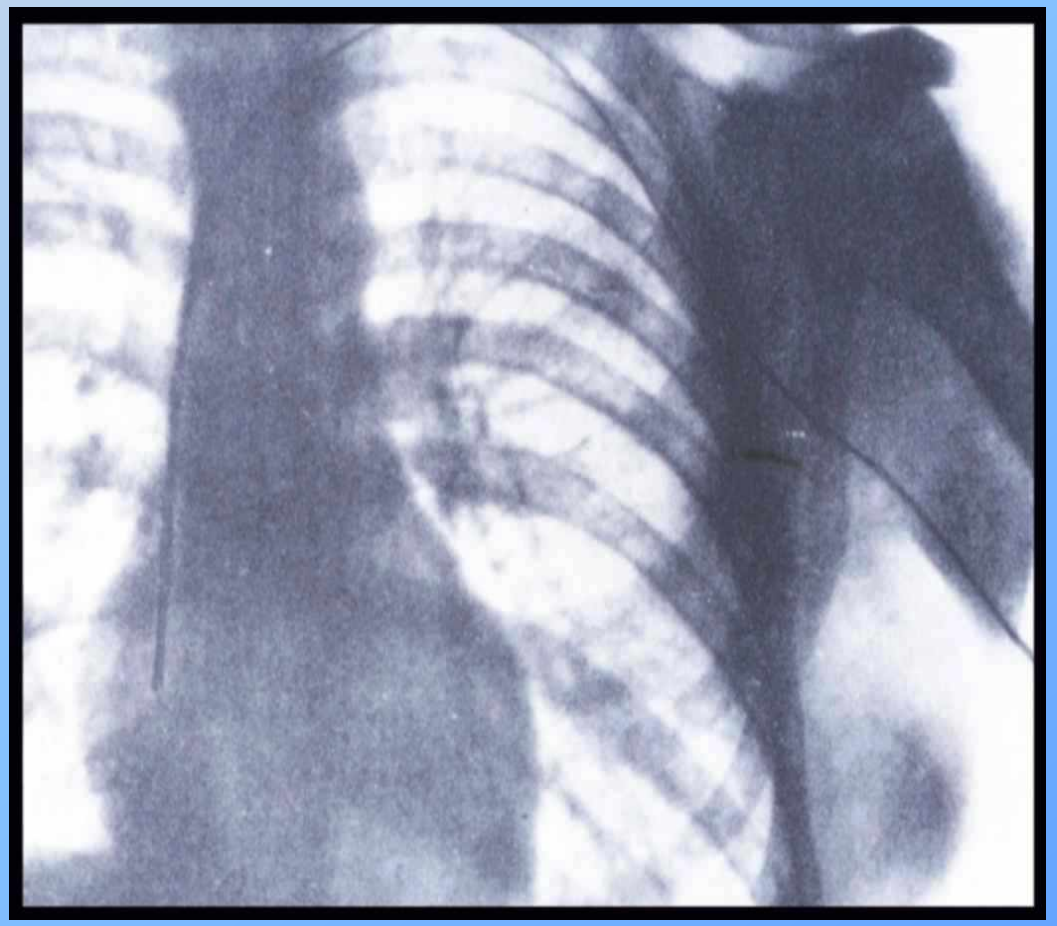
When he held his hand before the fluorescing screen, he was astonished to see the bones of his thumb and fingers. In 1901, Wilhelm Konrad Roentgen, a German experimental physicist, was awarded the first Nobel prize.

The Internal Structure of the Living Heart

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

"The internal structure of the living heart had been revealed for the first time..."

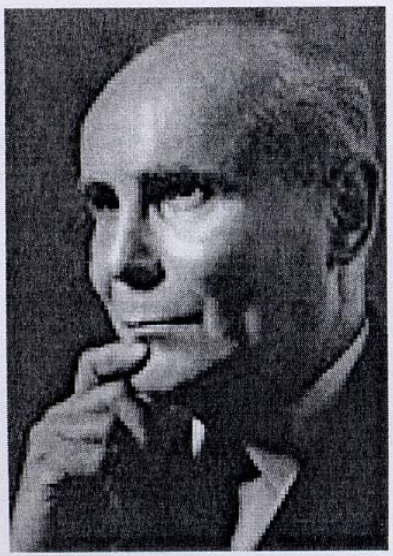
1929
Werner Forssman
The Cardiac Catheter



Prelude to the Nobel

In 1870, Fick and Grollman established in dogs a principle for measuring cardiac output. Andre F. Cournand and Dickenson W. Richards of New York used Forssmann's cardiac catheter to apply the Fick principle in human subjects.

The 1956 Nobel Prize in Physiology or Medicine was awarded to André F. Cournand, Werner Forssmann and Dickinson W. Richards *"for their discoveries concerning heart catheterization and pathological changes in the circulatory system".*

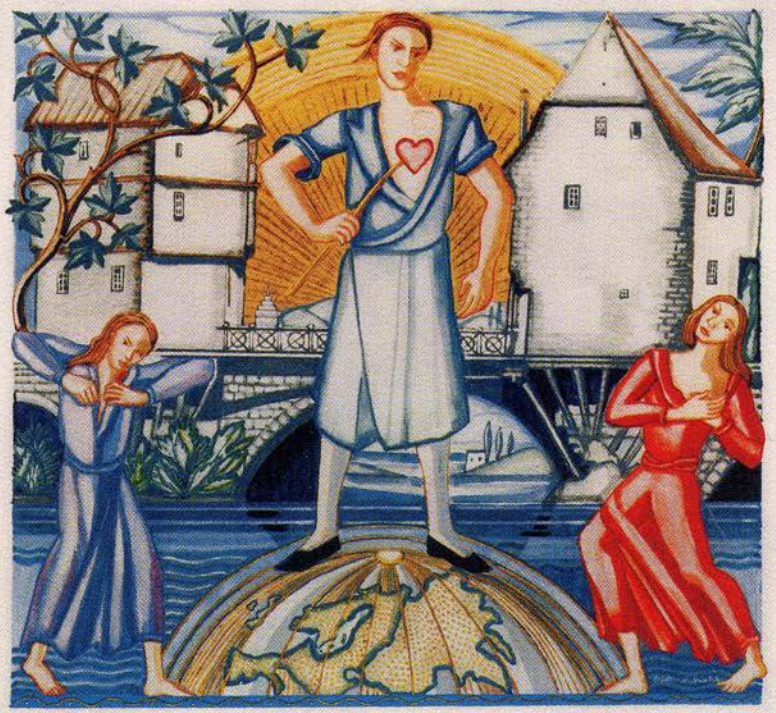


Andre Frederic Cournand

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

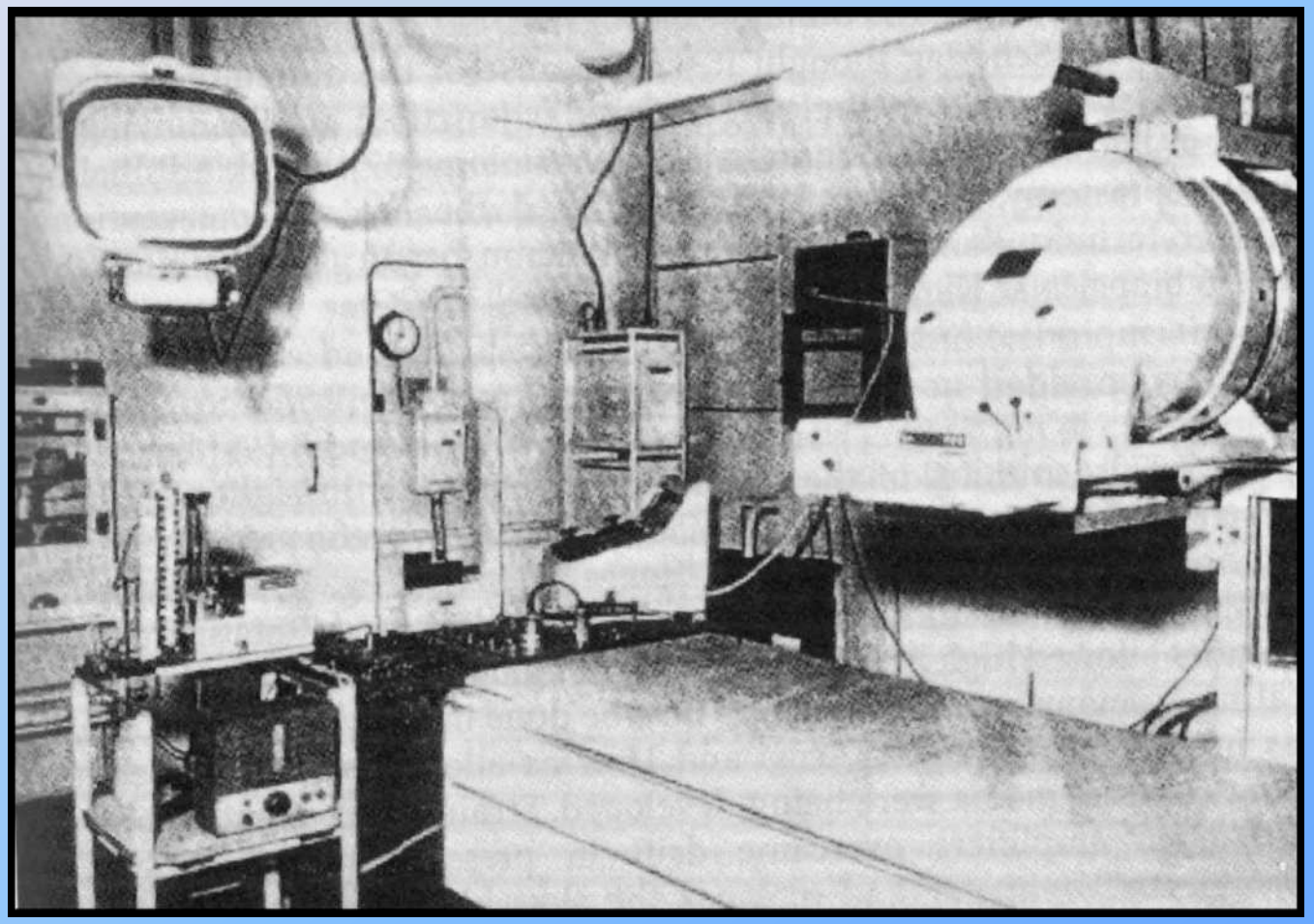
ANDRÉ COURNAND
WERNER FORSSMANN
OCH
DICKINSON W. RICHARDS

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

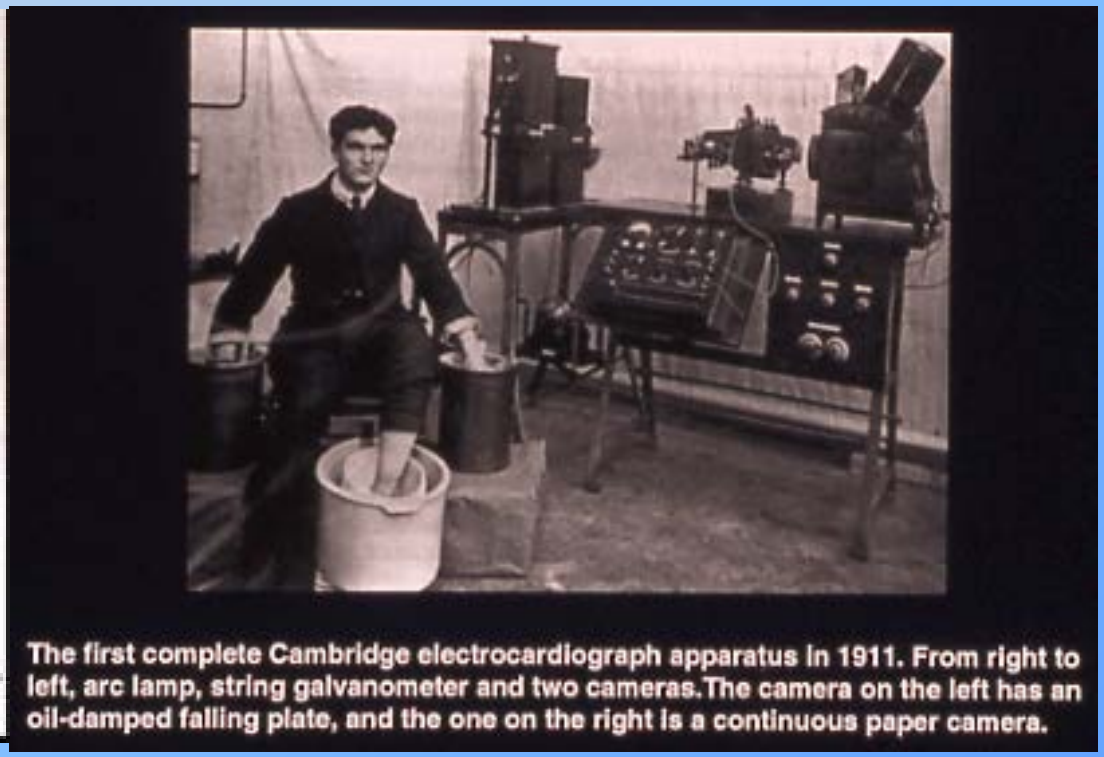
Anatomic 1906

While working in Ludwig Aschoff's laboratory in Marburg, Suano Tawara published his epoch-making monograph, *The Conduction System of the Mammalian Heart.*

Electrocardiographic 1924

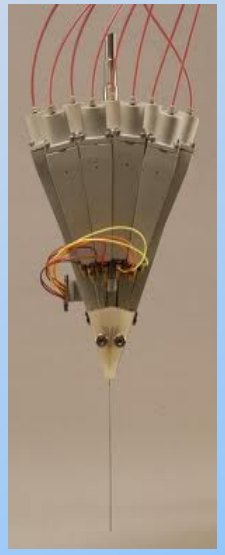
Willem Einthoven (1860-1927)

Nobel prize in physiology or medicine



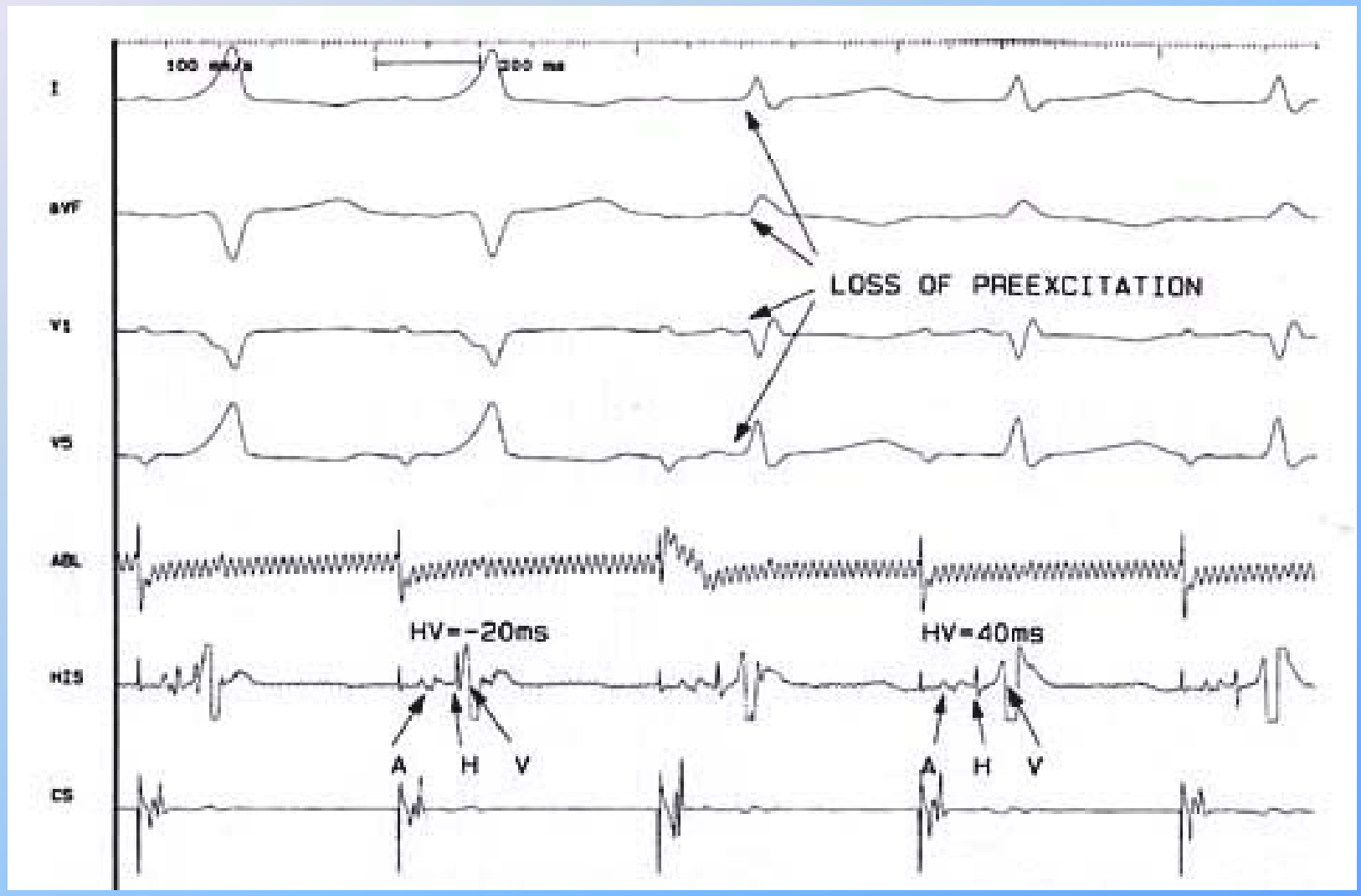
The first complete Cambridge electrocardiograph apparatus in 1911. From right to left, arc lamp, string galvanometer and two cameras.The camera on the left has an oil-damped falling plate, and the one on the right is a continuous paper camera.

Electrophysiologic 1949



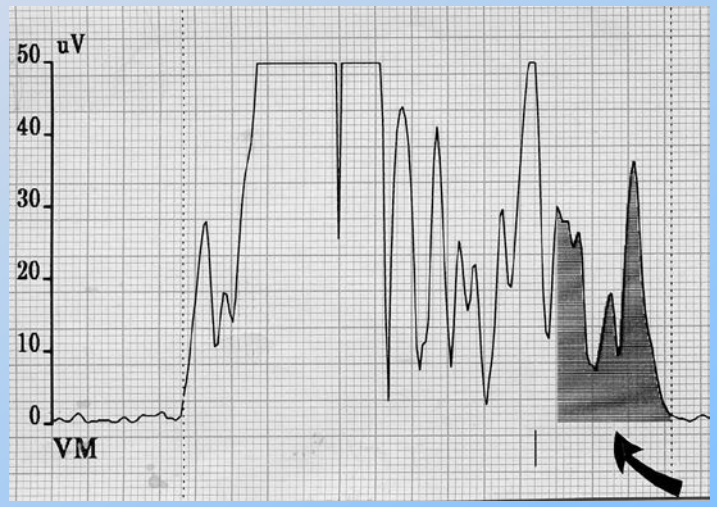
Electrophysiology had its inception in 1949 with Ling and Gerard's introduction of the microelectrode which has a tip that can be placed adjacent to or inside a cell for the purpose of recording electric potentials.

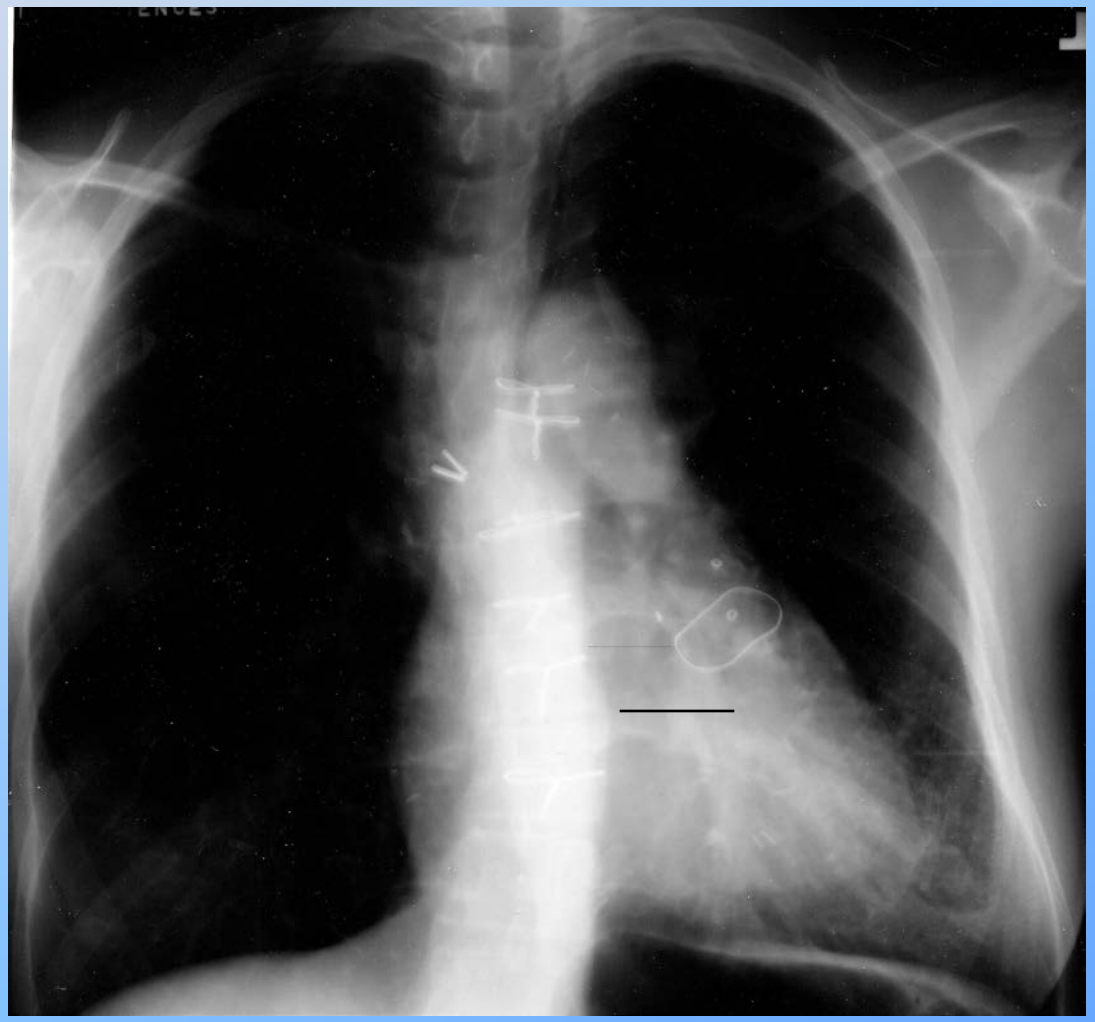
Accessory Pathways
Radiofrequency Ablation



A Slow Conduction Substrate

The Signal Averaged Electrocardiogram

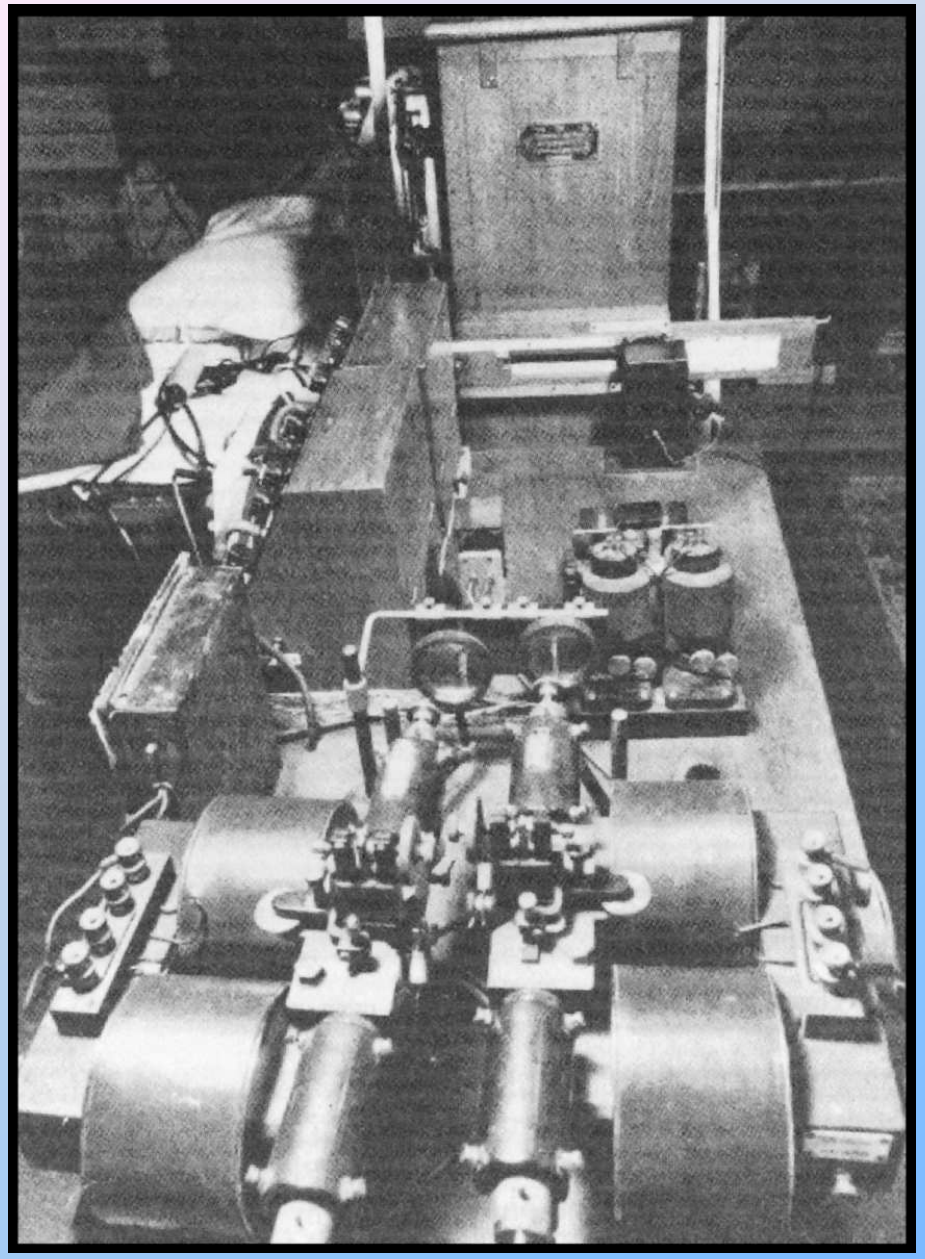




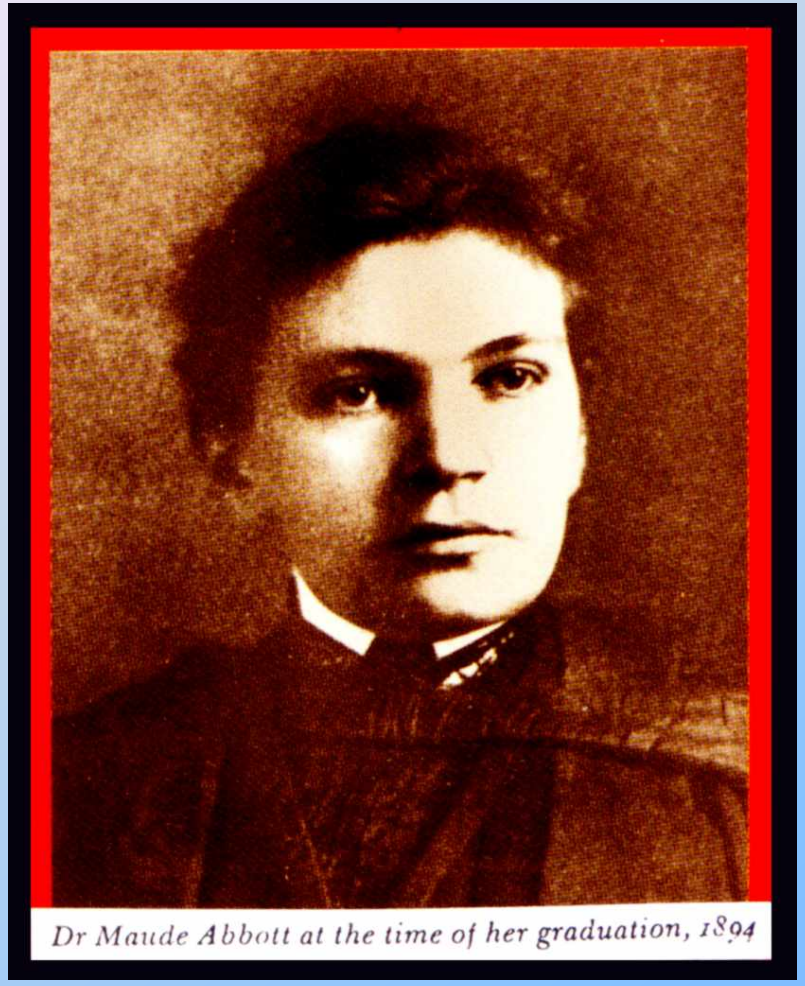
The Audible Language of the Heart

PHONOCARDIOGRAPHY

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



Leatham's phonocardiograph consisted of two Cambridge string and two mirror galvanometers. (Welcome Museum, London.)



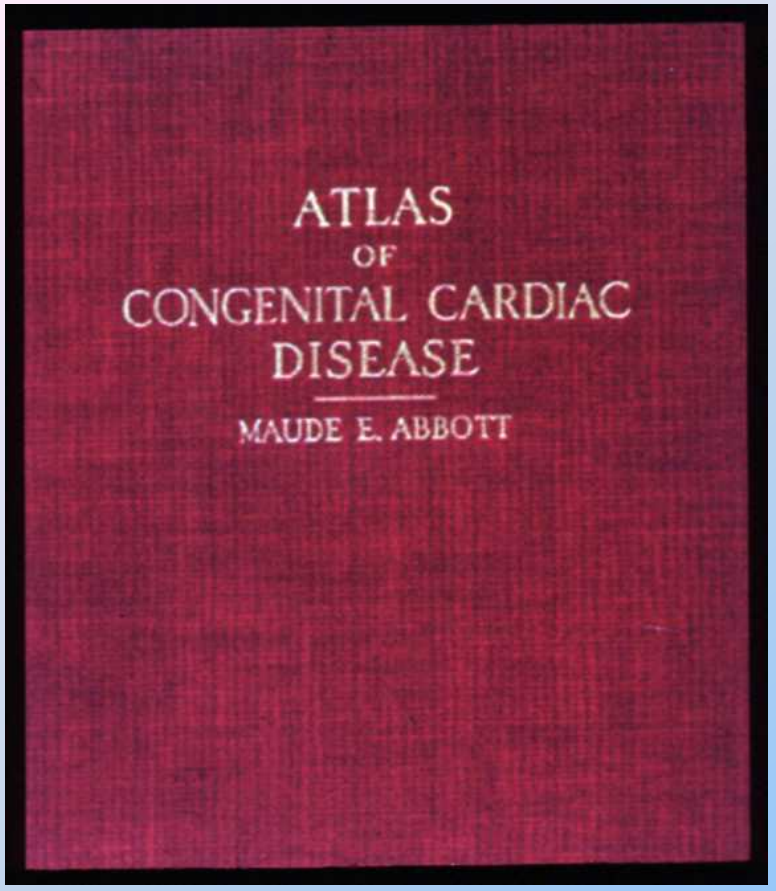
Maude Abbott

Helen Taussig

Before World War II, congenital malformations of the heart were regarded as *hopeless futilities*, an occupation 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. ***Helen Taussig*** was advised to occupy herself with the *hopeless futilities* in the children's clinic at Hopkins. Congenital heart disease in adults was virtually unknown.

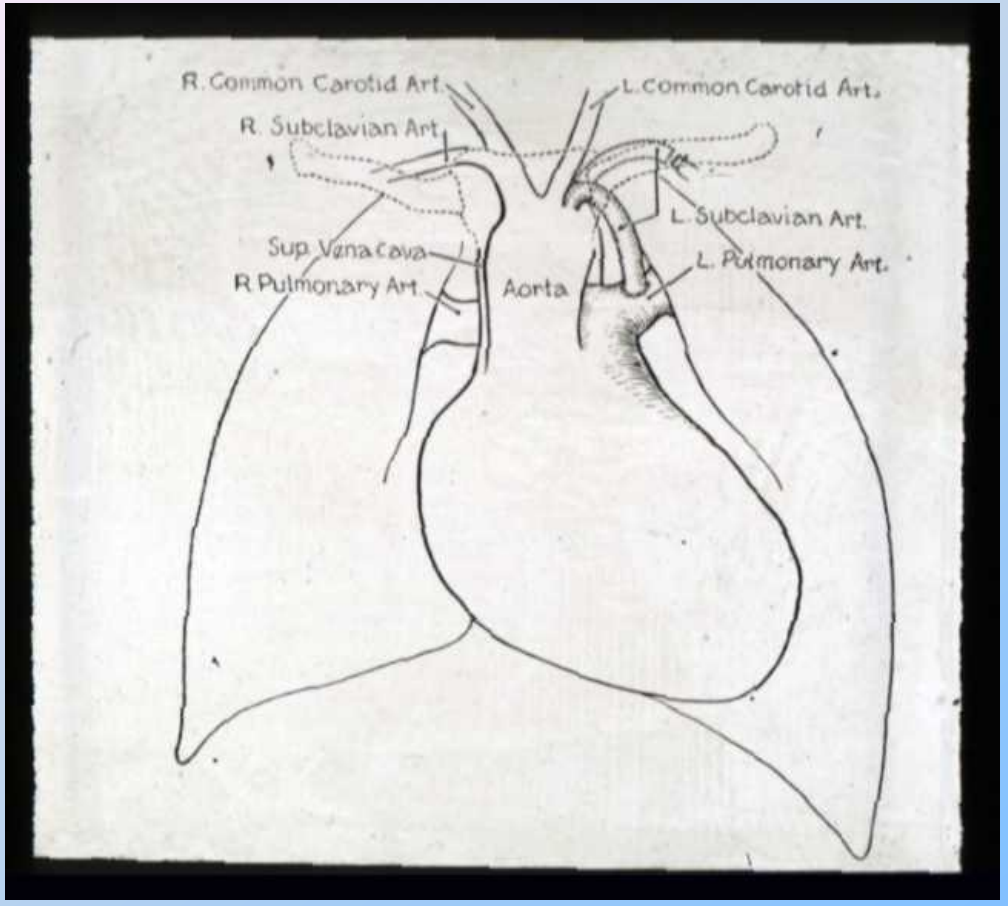
But now, approximately 85% of infants with congenital heart disease in developed countries reach adulthood.

The hopeless futilities have come of age.

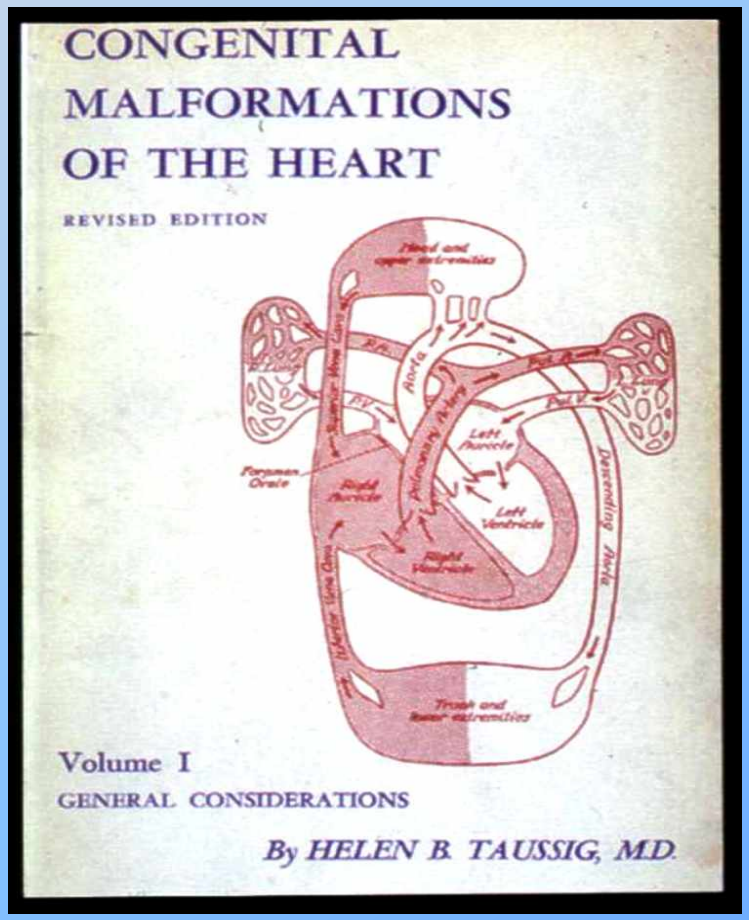


1936

The Osler Library
McGill



The Blalock Taussig Operation 1944



Taussig's Book 1947

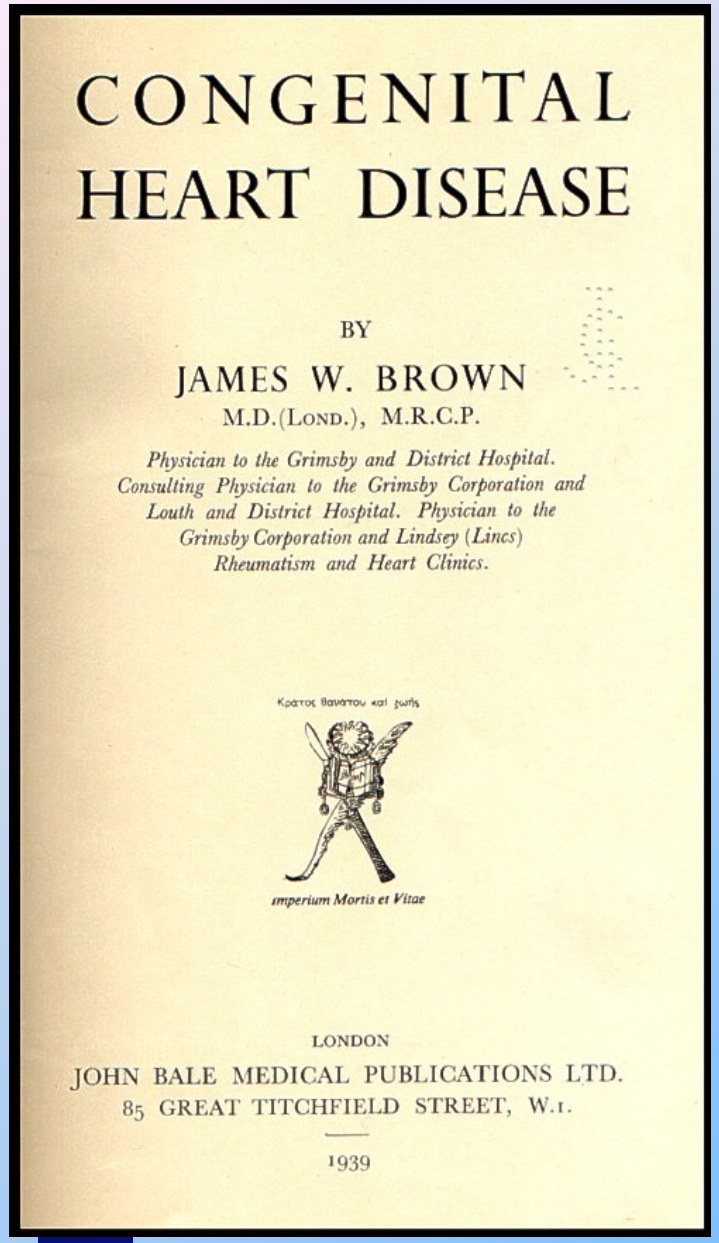
The Coming of Age

I shall 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.

Preface to James W. Brown


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

It has been 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 devoted to the study of heart disease in children has made it possible to observe more than 350 cases of congenital heart disease. These observations form the basis of this work.

First Edition 1939

First Steps

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

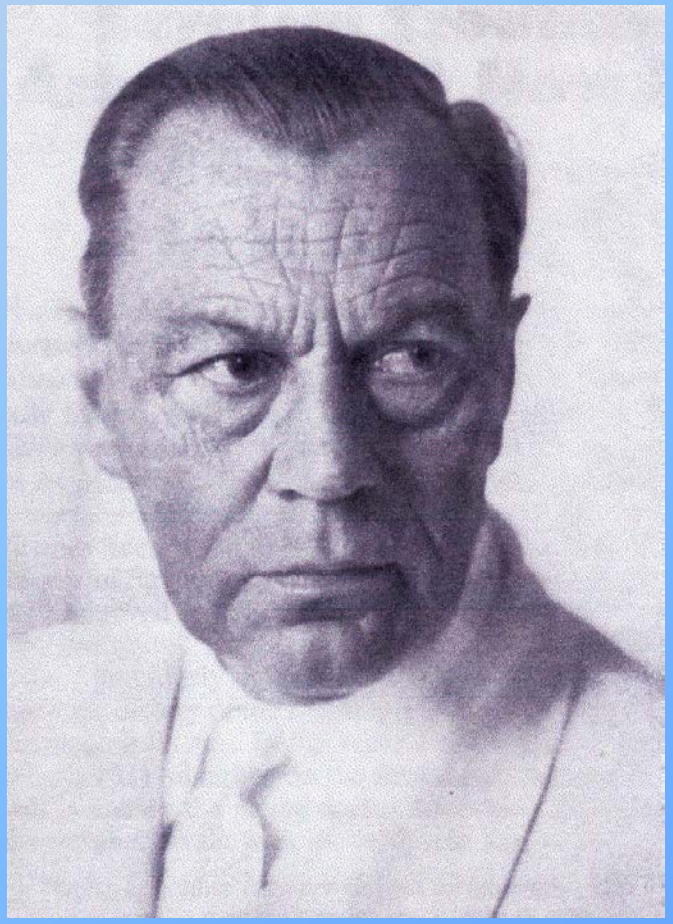
SURGICAL LIGATION OF A PATENT DUCTUS ARTERIOSUS: REPORT OF FIRST SUCCESSFUL CASE

R.E. GROSS

and J. P. HUBBARD

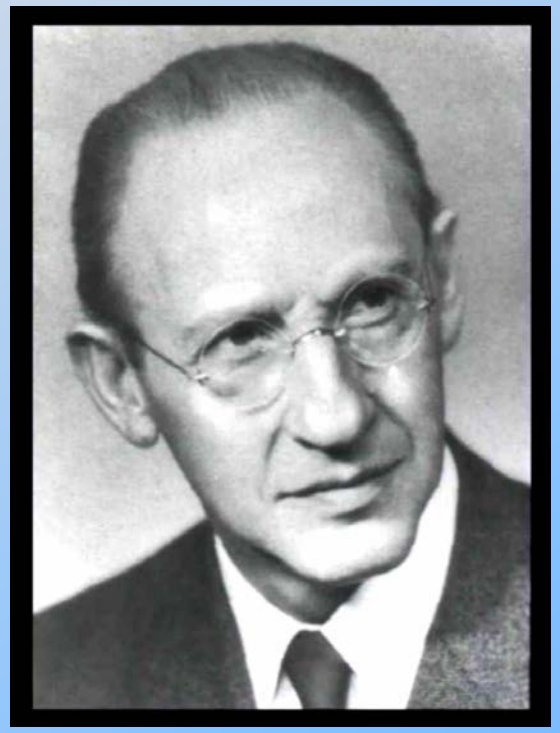
1939

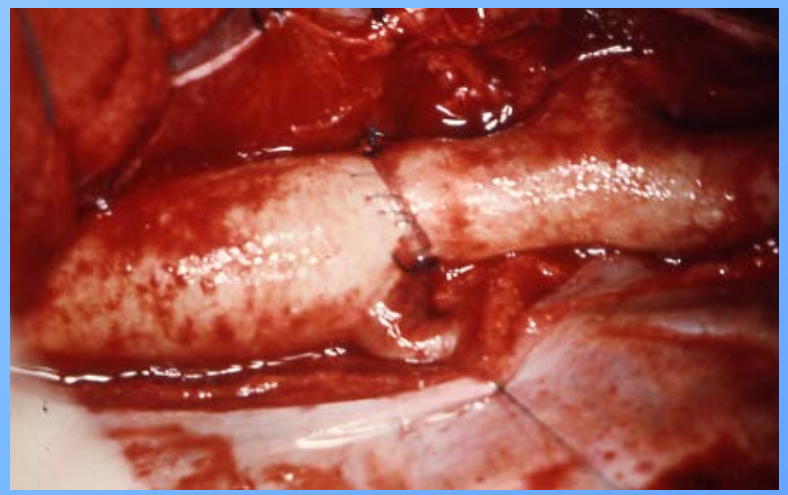
In 1938, E.K. Frey, a surgeon in Dusseldorf, performed the first successful ligation of a patent ductus in a 14 year old boy. Hospital documents were destroyed during the second world war. The case was not reported.



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 the experimental work of Gross and Hufnagel, 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.





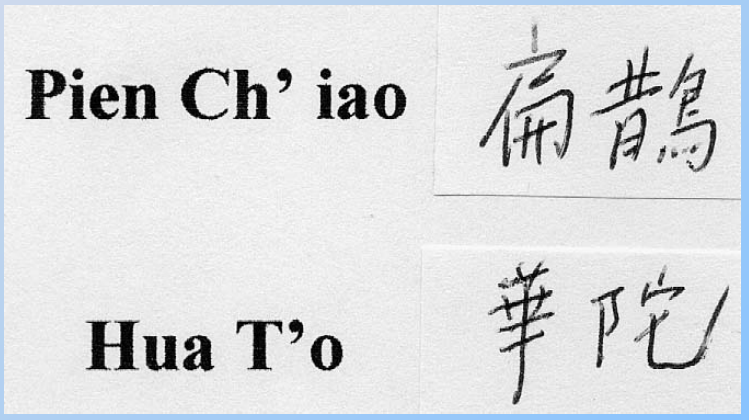
The Pivotal Role of Anesthesia

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 in the United States and ranks among America's greatest contributions to medicine.

The Genesis of Anesthesia was *Genesis*

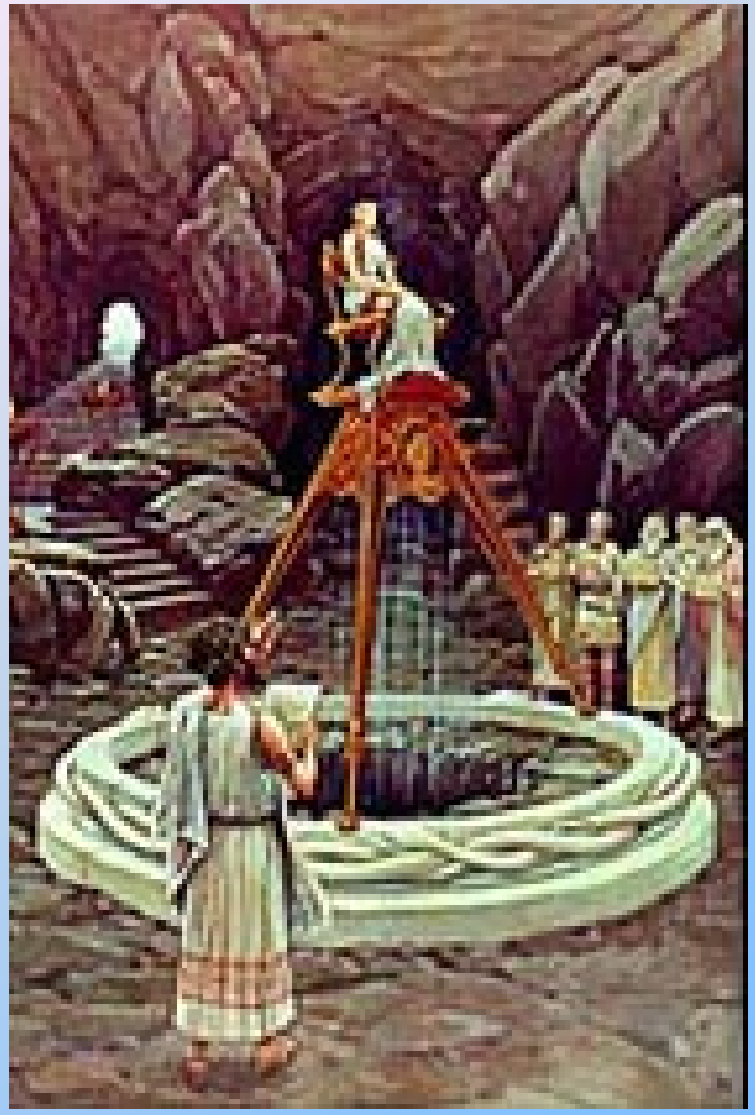
And the Lord God caused a ***deep sleep*** to fall upon Adam, and he slept: and he took one of his ribs, and closed up the flesh thereof.

Chinese Medical History



The writings of Hua T'o on surgery and anesthesia date from 190 AD.

Temple of Apollo in the Sanctuary of Delphi



The Delphic Oracle answered questions while in a trance-like state because her tripod was directly above a geographic fault from which arose fumes of ethylene, an anesthetic.

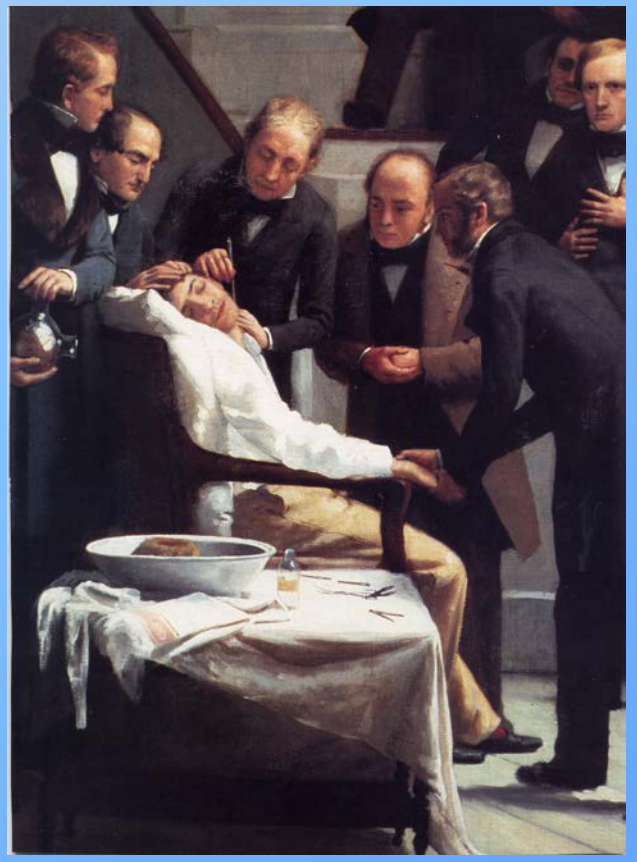
A Country Doctor

Ether was first used in surgery in March 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 temporary but complete insensibility to pain.***

Long's bill was $2 for the operation and 25 cents for the ether.

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. In the next month, ether was used in London. In the succeeding year ether became widespread throughout Europe.



Anesthetic Device Nineteenth Century. Science Museum, Oxford, UK

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.

Alfred Blalock

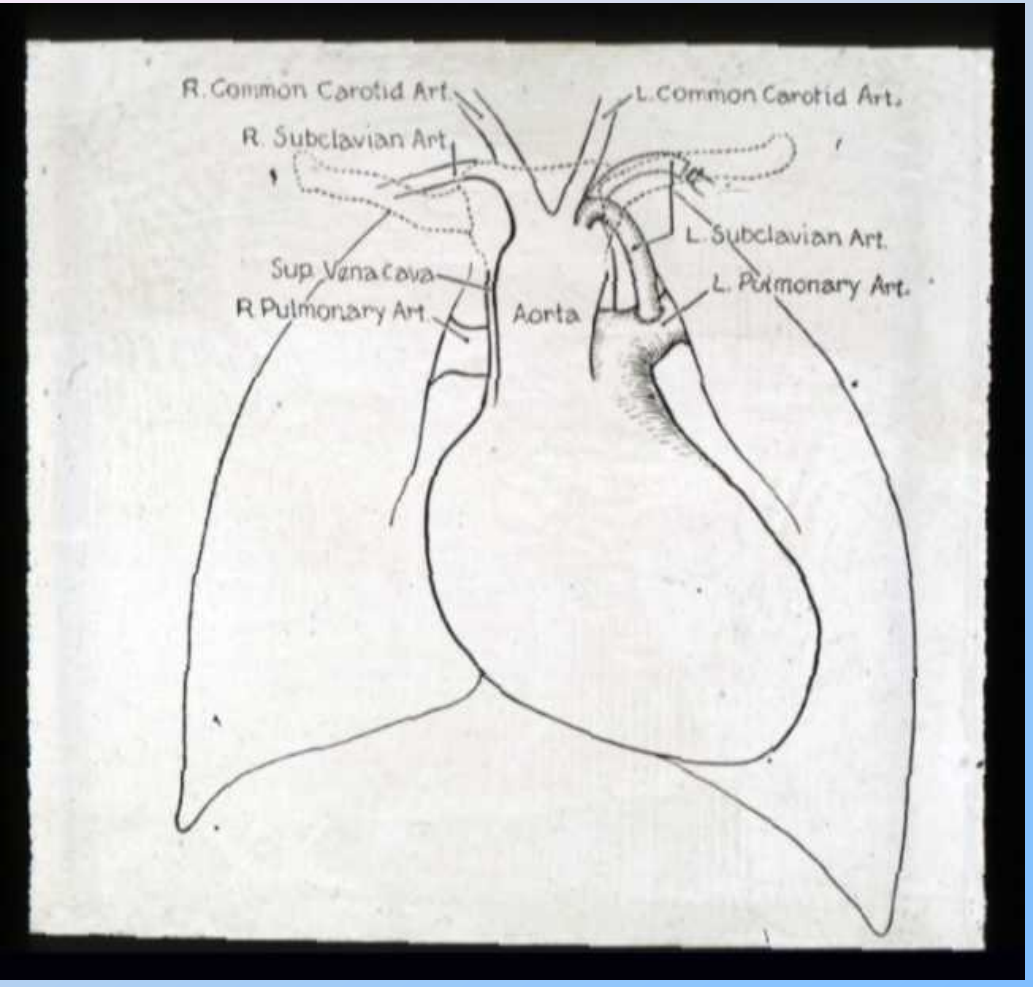
Helen Taussig

The Blalock/Taussig Shunt

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

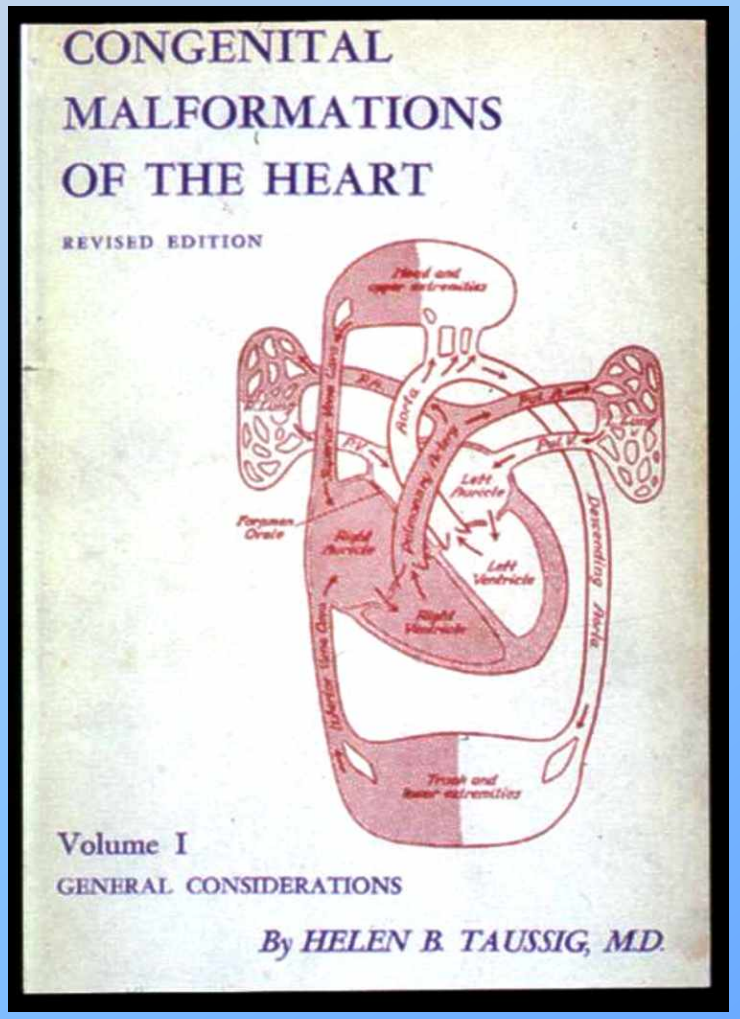
Merel Harmel

Eileen Saxon



A Landmark Operation

1944



1947

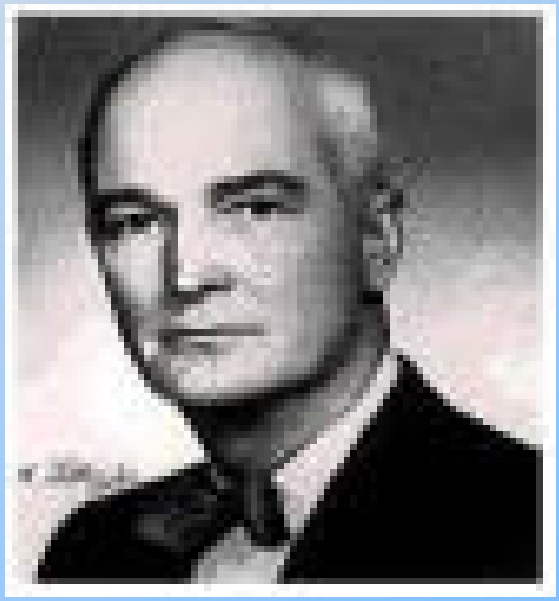
The Gibbon Pump

"…the patient's life might be saved if the blue blood in her veins could be withdrawn into an extracorporeal blood circuit, exposed to an atmosphere of oxygen, and then returned to the patient by way of a systemic artery."

John H. Gibbon

Surgical Fellow

Massachusetts General Hospital1931

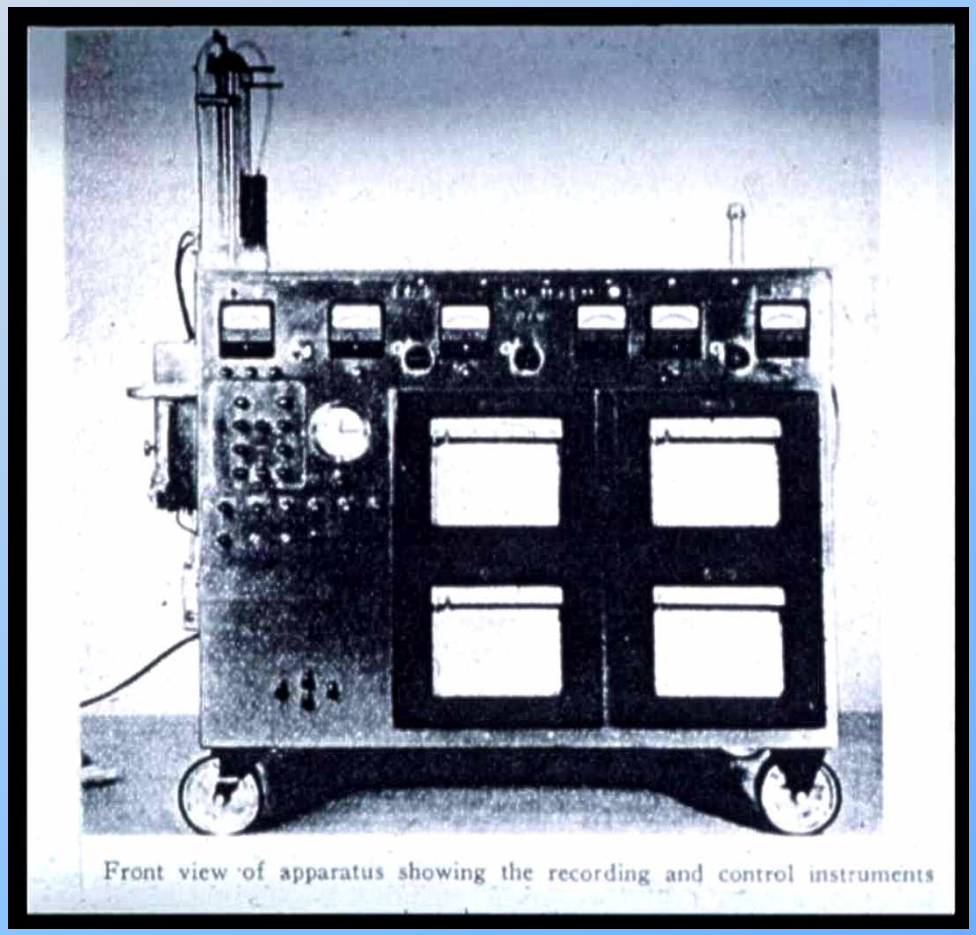


Minnesota Med. 37:171,1954

APPLICATION OF A MECHANICAL HEART AND LUNG APPARATUS TO CARDIAC SURGERY

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

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.



Front view of apparatus showing the recording and control instruments

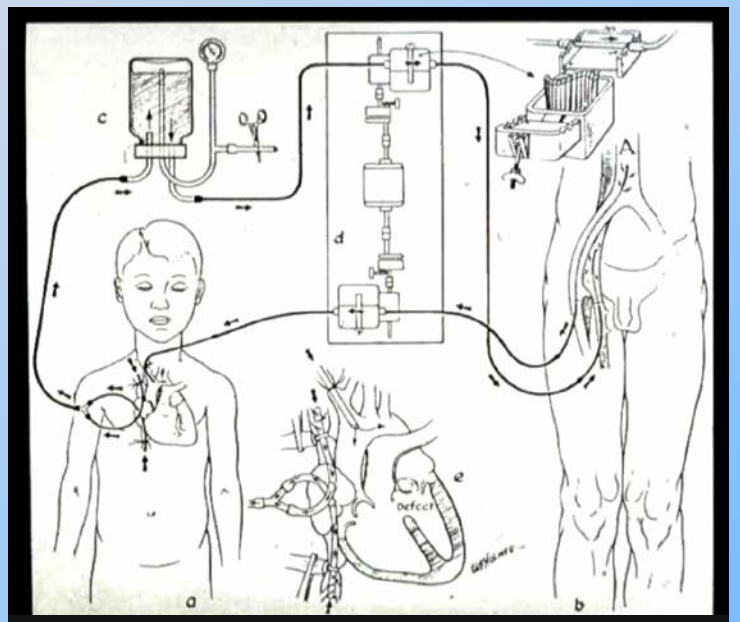
The Gibbon Pump Oxygenator

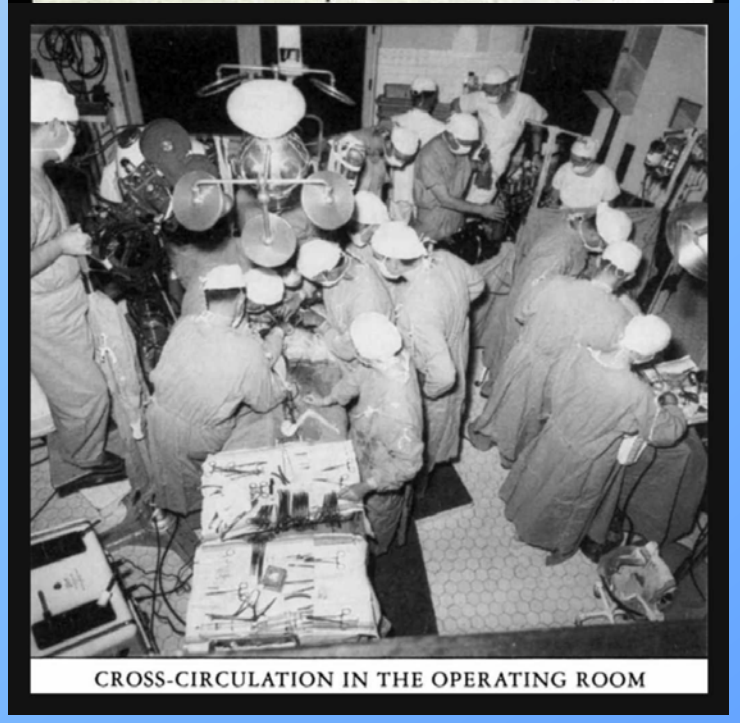
2012

HEART AND LUNG APPARATUS

1955

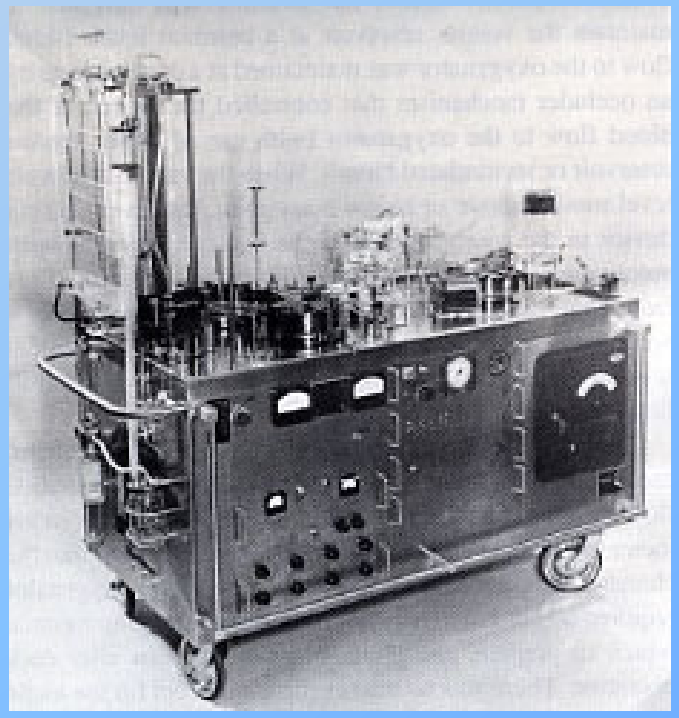
C. Walton Lillehei



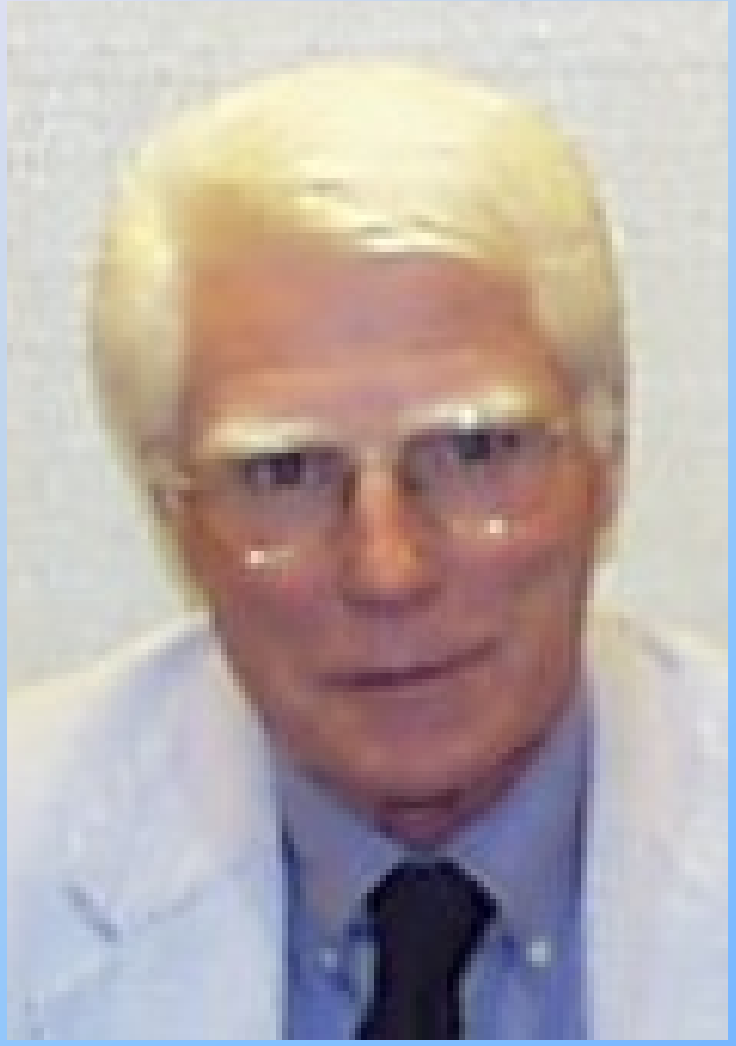


CROSS-CIRCULATION IN THE OPERATING ROOM

John W. Kirklin



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

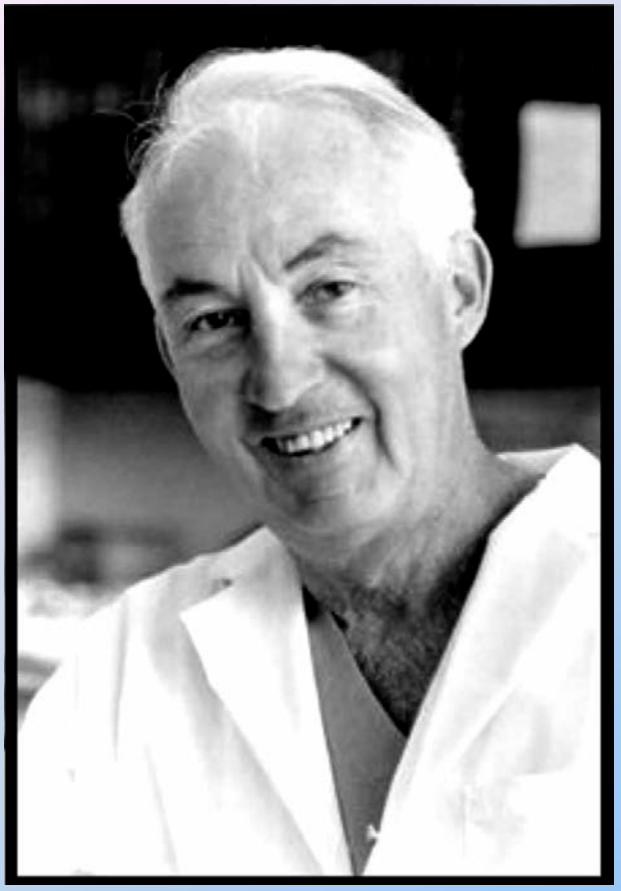


Aldo Casteneda, MD,PhD
Cardiac Surgery in the Neonate and Infant 1994

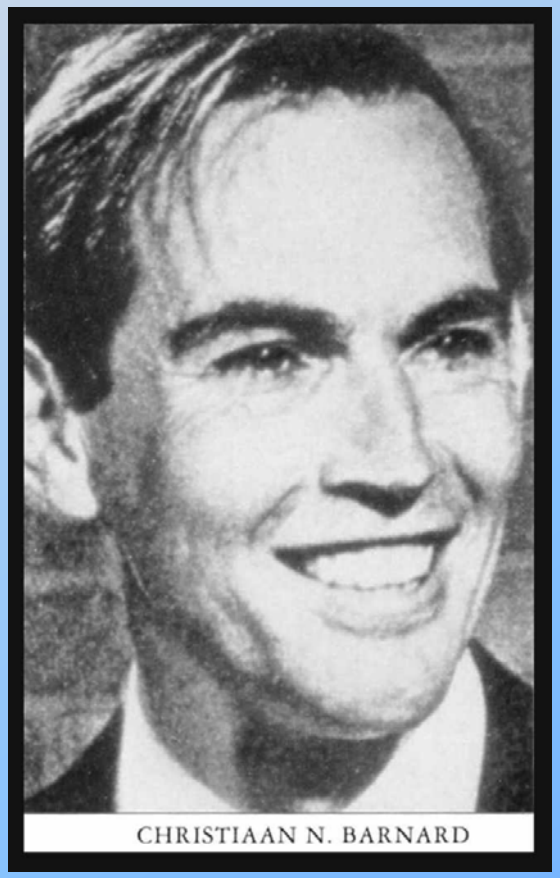
The First Cardiac Transplantation?

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---the first successful orthotopic cardiac transplantation.

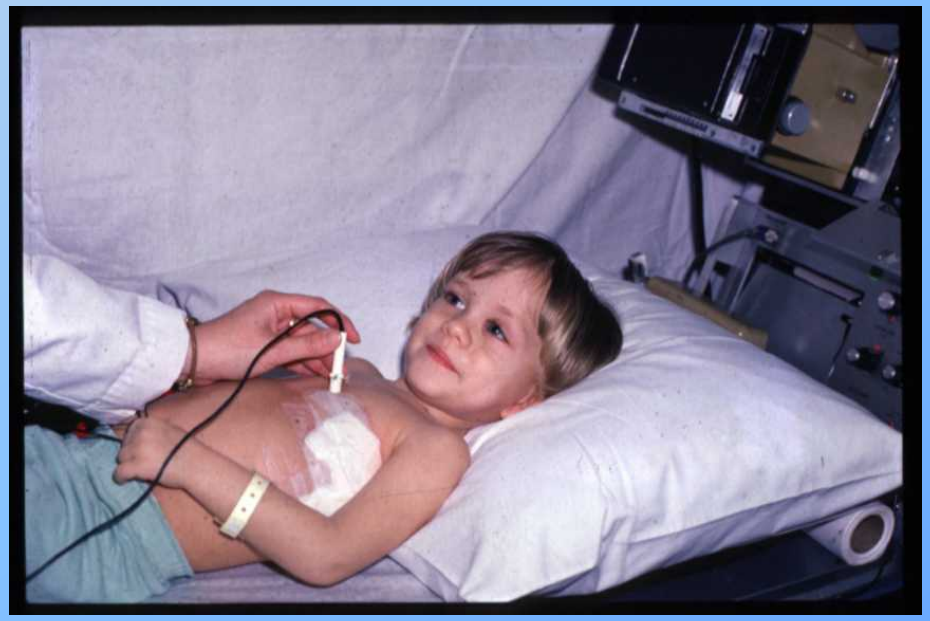


1966 — Christian Barnard---the first human cardiac transplantation.

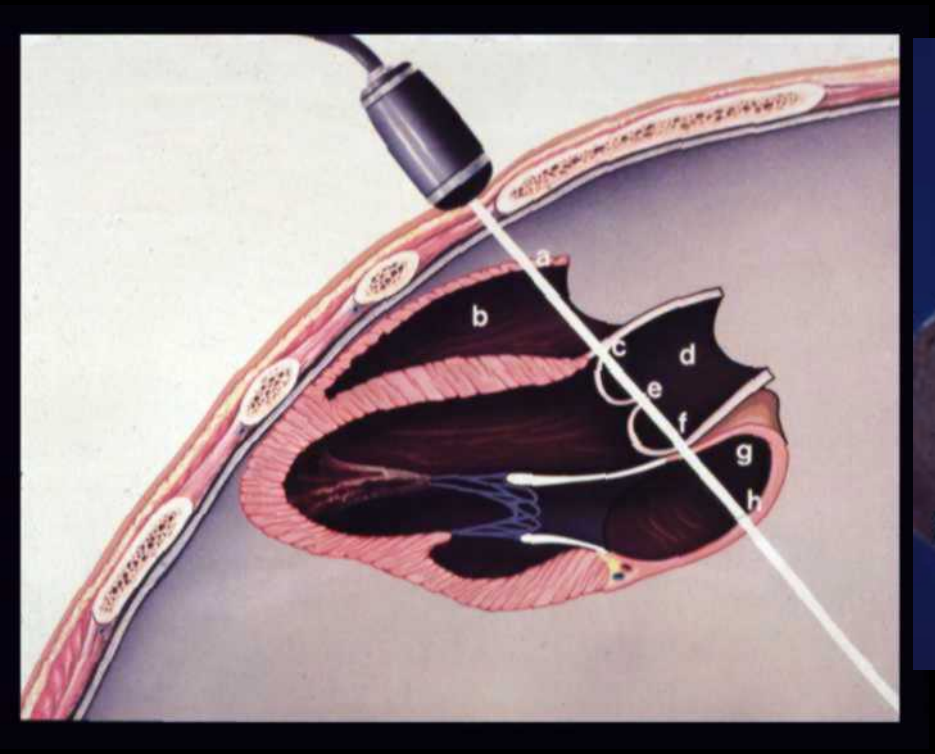
Echocardiocargraphy

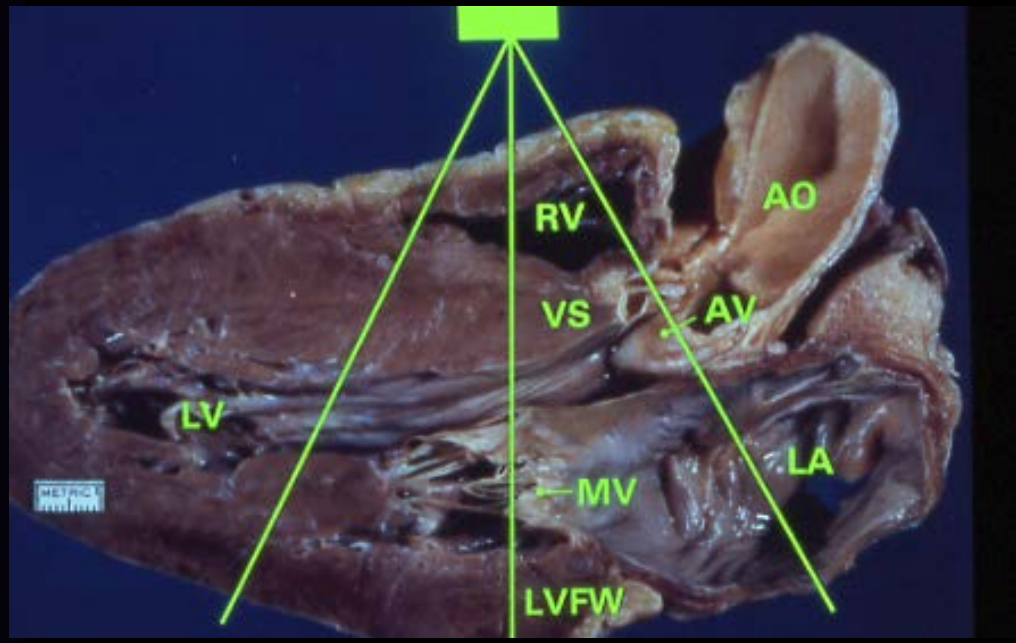
The Diagnostic Pathway to the Promised Land

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

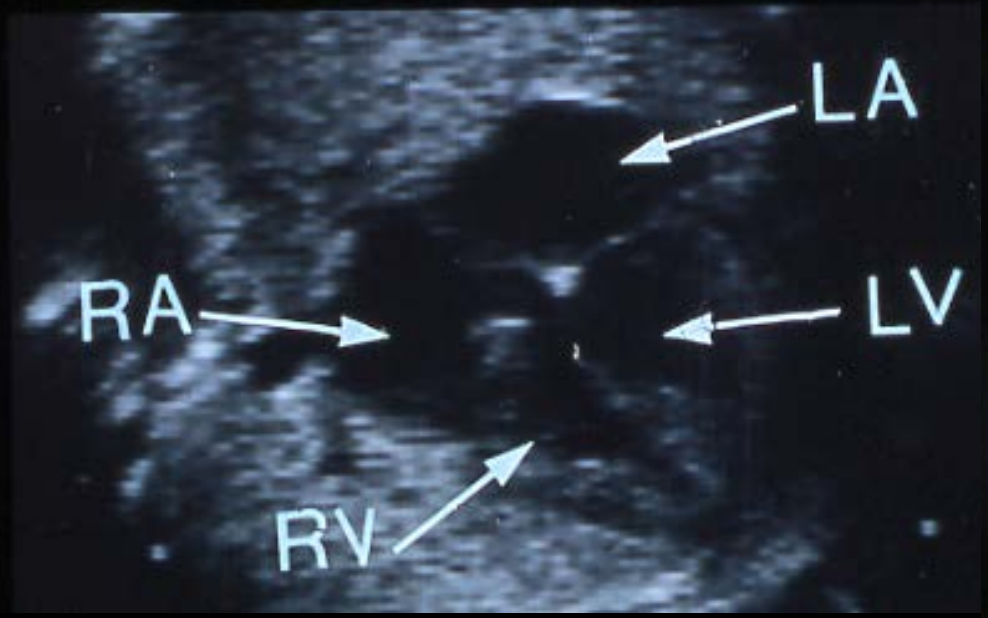


Transthoracic

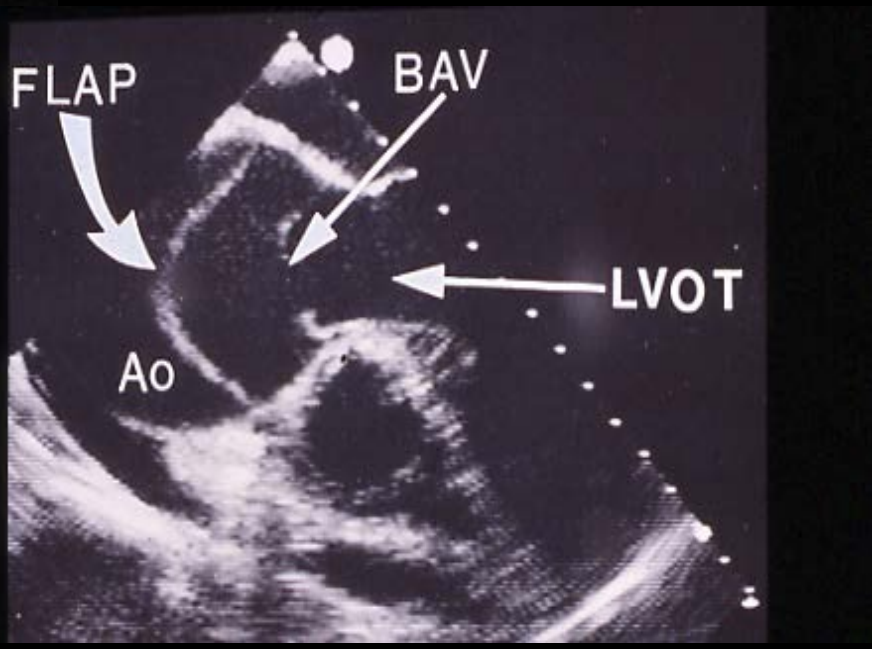




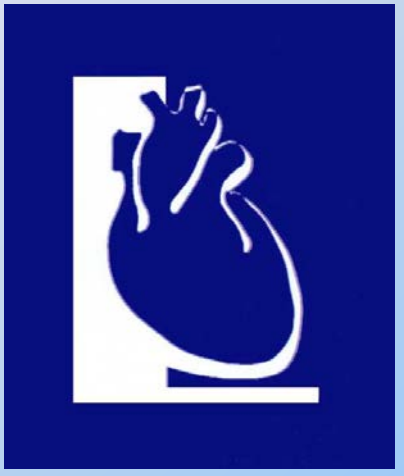
Fetal



Transesophageal



Gadolinium-Enhanced Magnetic Resonance Image



Cardiac Catherization

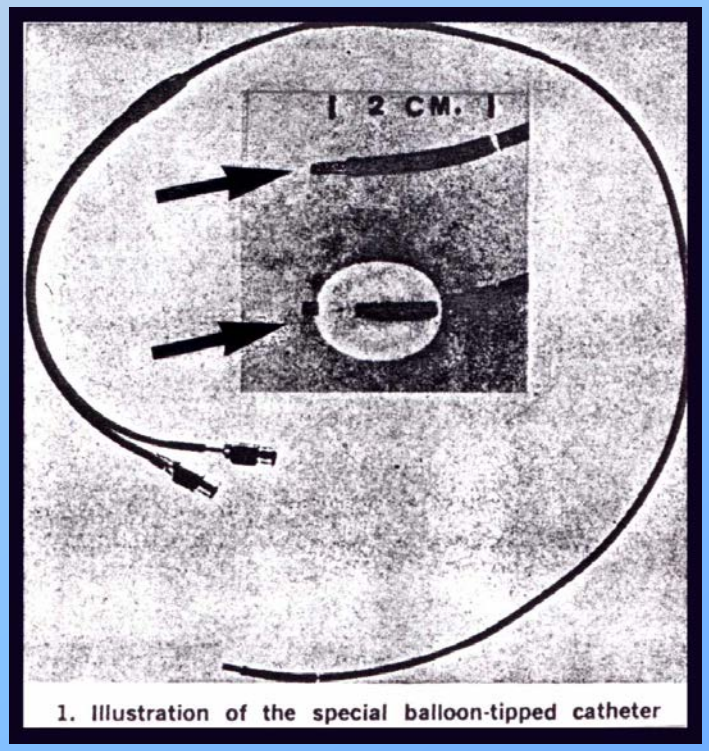
A Therapeutic Intervention

Am. Med. Assoc. J. 1966

Creation of an Atrial Septal Defect Without Thoracotomy

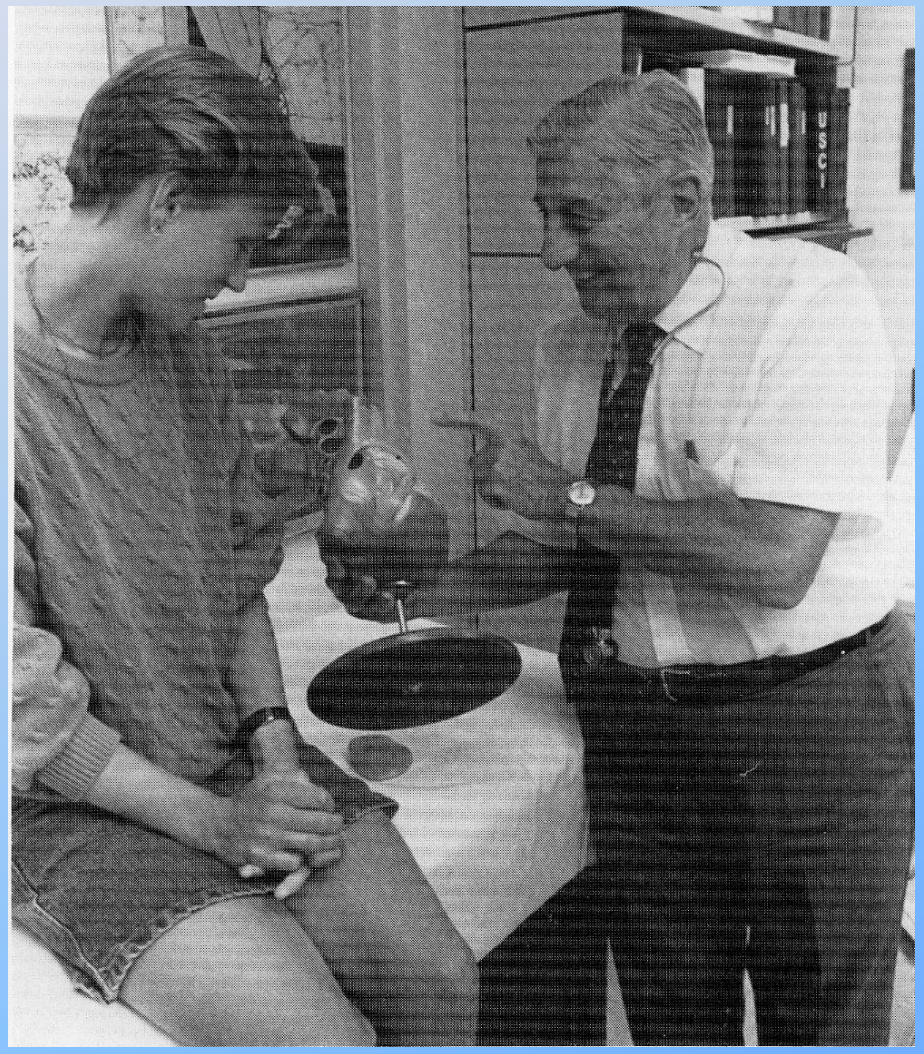
A Palliative Approach to Complete Transposition of the Great Arteries

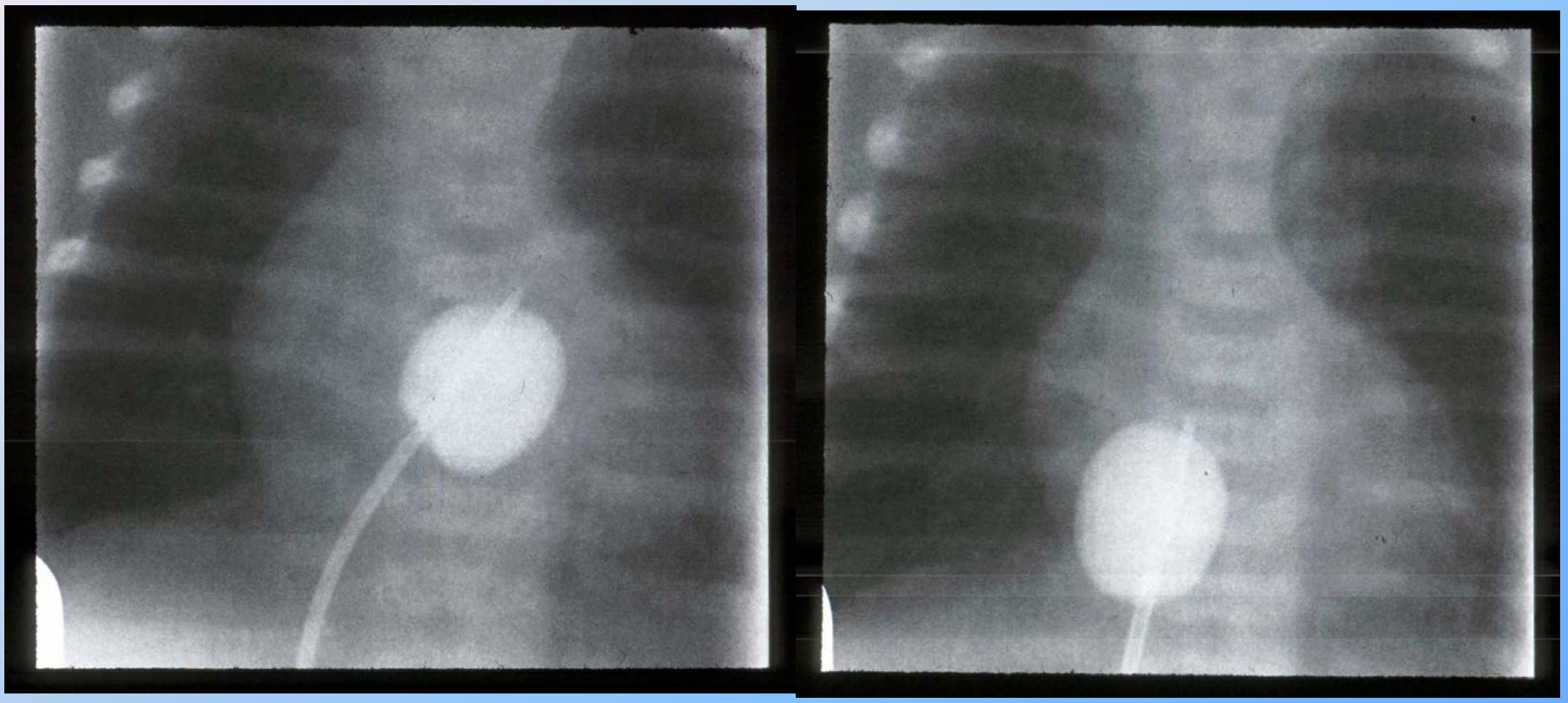
William J. Rashkind, MD, and William W. Miller, MD



1. Illustration of the special balloon-tipped catheter

William J. Rashkind

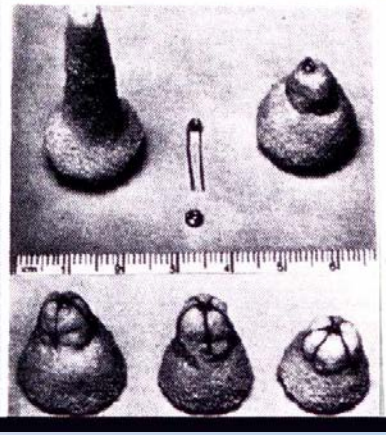




"Success depends on the jerk at the end of the catheter."

WJR

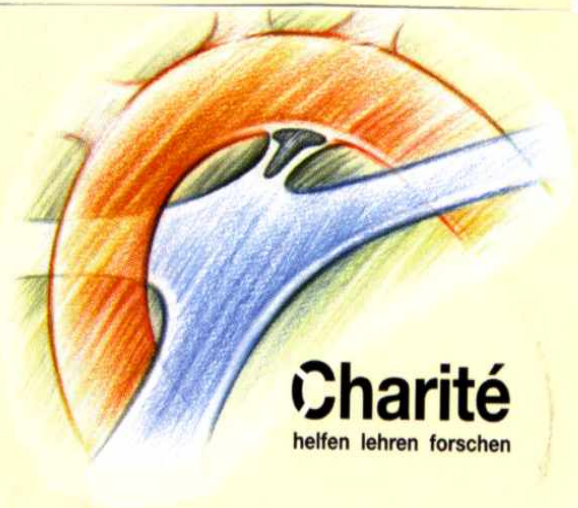
The first publication about the interventional closure of patent ductus arteriosus in 1967.



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

30 YEARS

interventional closure of patent ductus arteriosus



Universitätsklinikum Charité
Interdisciplinary Symposium Berlin,
11.-12.10.1996

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 assume 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 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.

Tertiary Care for Adults With CHD

Specialized facilities for the comprehensive care of adults with congenital heart disease offer services difficult if not impossible to duplicate.

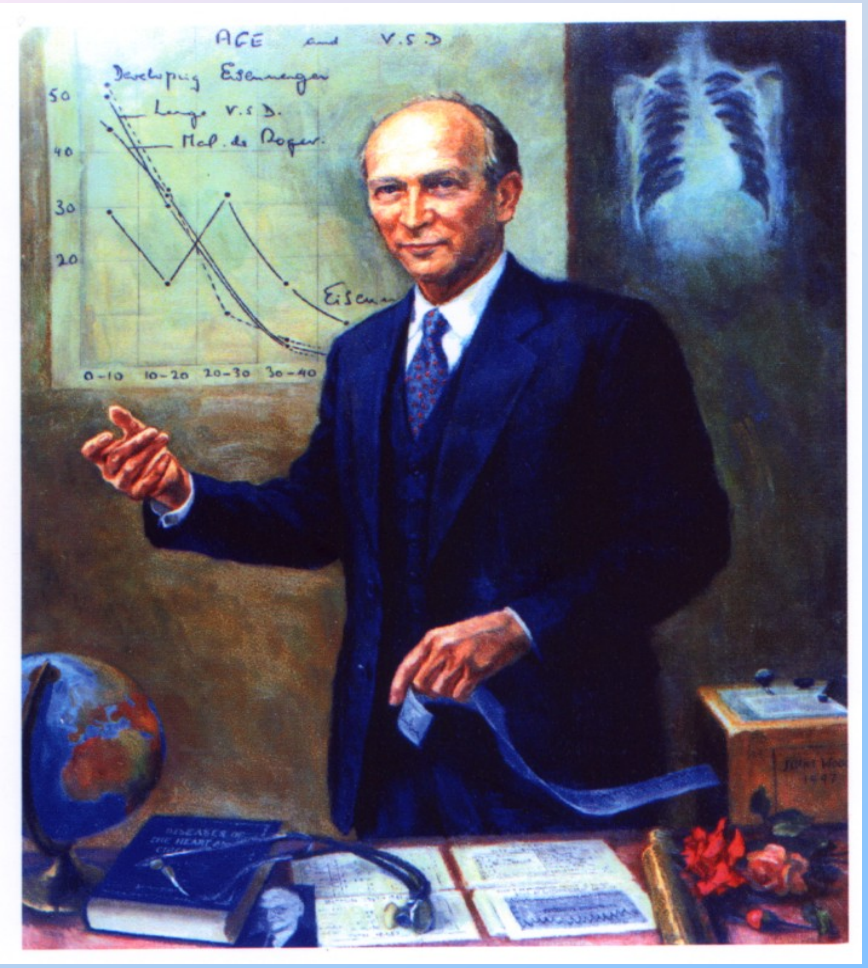
Personal Reminiscences

Turning the efforts of many years into an hourglass

London 1954


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

The 1950's



Portrait by Wood's daughter Juliet

DISEASES OF THE HEART AND CIRCULATION

by

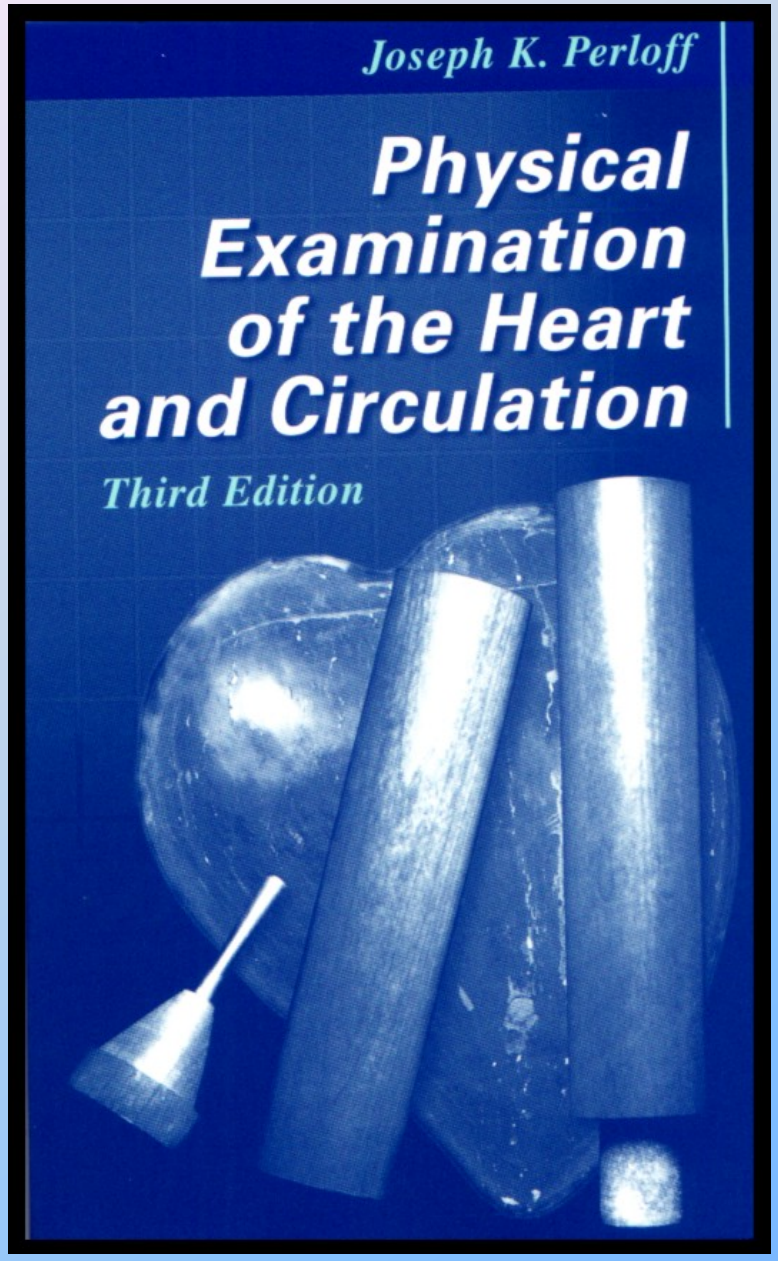
PAUL WOOD, O.B.E.

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

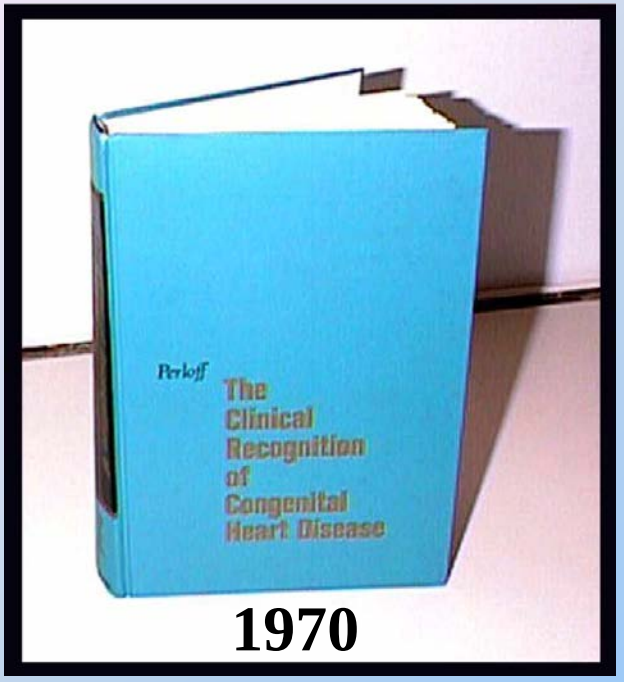
Director, Institute of Cardiology, London
Physician, National Heart Hospital
Physician in charge of the Cardiac Department, Brompton Hospital

CHAPTER II. PHYSICAL SIGNS

CHAPTER VIII. CONGENITAL HEART DISEASE



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



1970

This book ***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

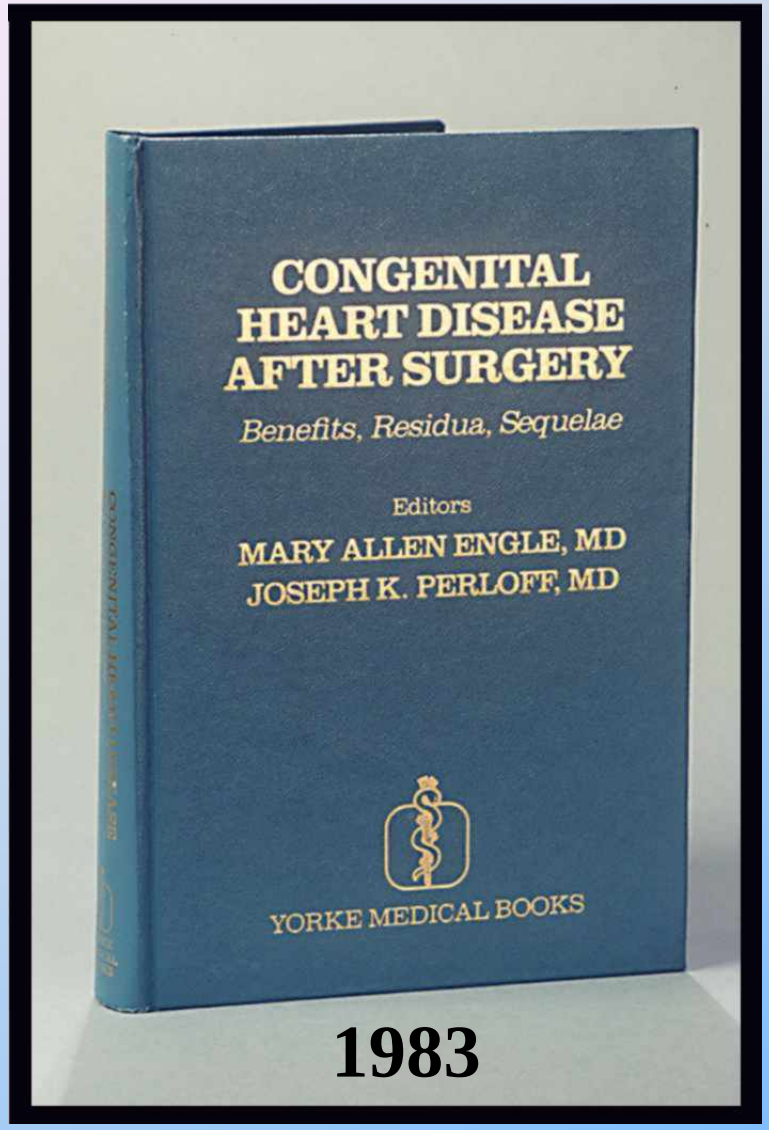
Forty Years Ago

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



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

Mary Allen Engle

Joseph K. Perloff

Congenital Heart Disease in Adults The Future of Children

The Infant

The Postoperative Adult

Congenital Heart Disease Approximate Worldwide Incidence

- **Moderately Severe Malformations--3/1000 live births.**
- **Severe Malformations----2.5-3/1000 live births.**
- **Collective incidence---Approximately 0.6% of live births.**

- **A bicuspid aortic valve occurs in about 2% of live births**

Adult Congenital Heart Disease in the United States

About 1,000,000.

About 20,000 open operations per year.

Annual increase approximately 5% per year.

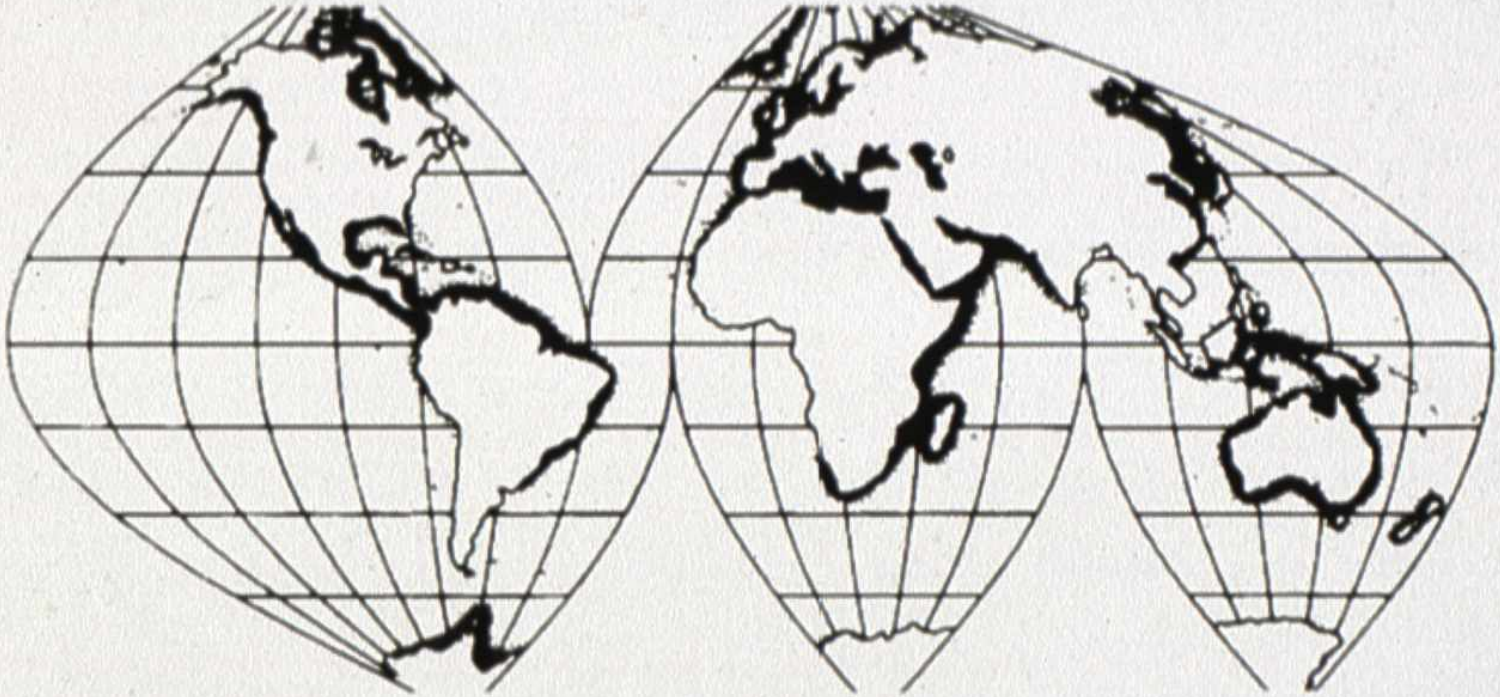
Currently, there are more adults with CHD in the US than there are infants and children.

Worldwide Incidence of Congenital Heart Disease

1.5 million new cases per year.

Approximately **85%** of afflicted infants in developed countries now reach adulthood

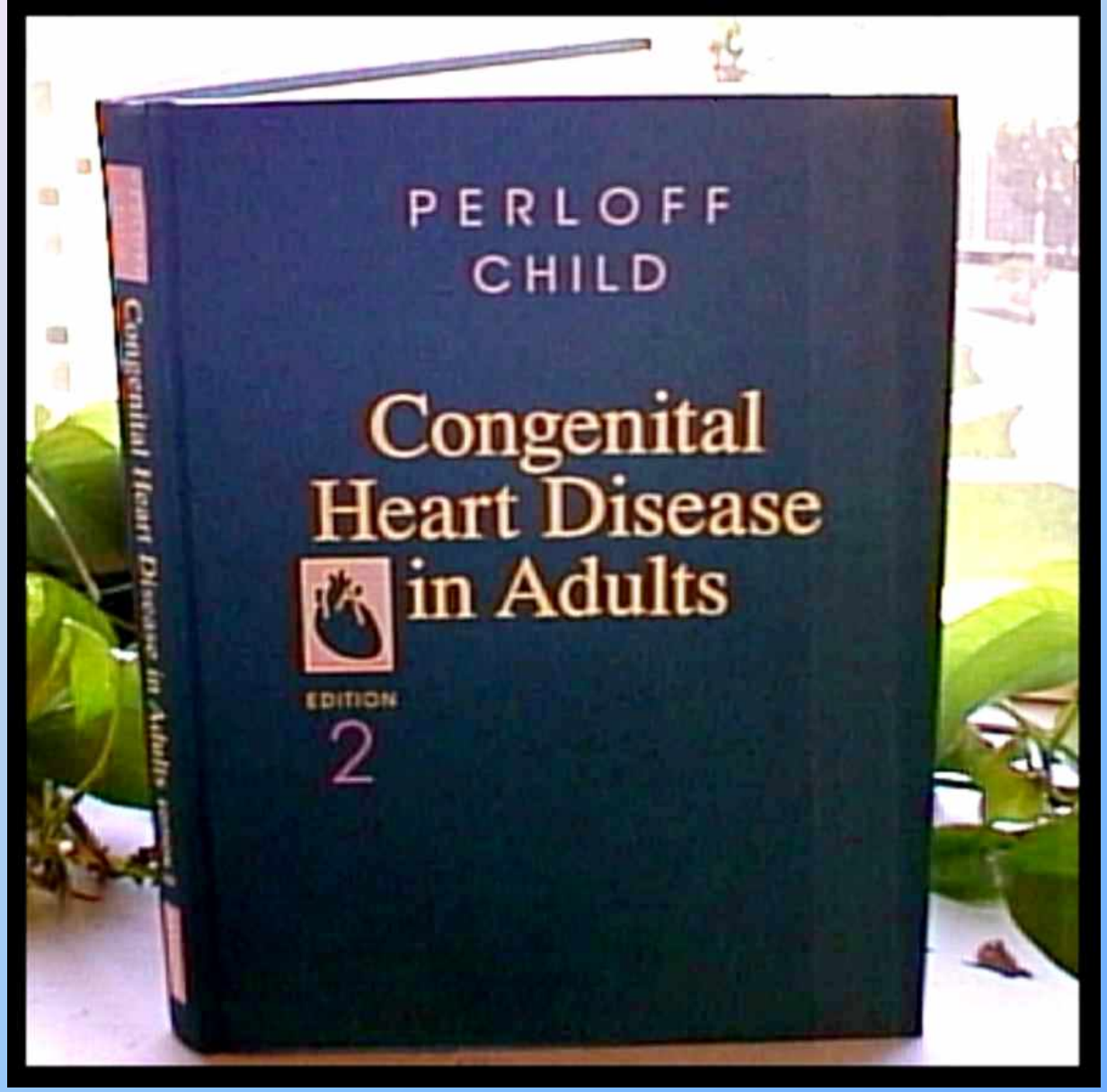
International Society for



Adult Congenital Cardiac Disease

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 2012
The UCLA Hospital

Ahmanson/UCLA Adult Congenital Heart Disease Center

EPILOGUE

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