Longevity In Unoperated Congenital Heart Disease

The Ahmanson/UCLA Adult Congenital Heart Disease Center
Unoperated and Untreated
The First Account

ATLAS
OF
CONGENITAL CARDIAC DISEASE
MAUDE E. ABBOTT
Natural and Unnatural

Natural history is not synonymous with unoperated. Surgeons are not perpetrators of the unnatural.
Gaussian Distribution is a statistical article of faith. However, it is the rarity at the far end of the distribution curve that provides insight into unoperated survival.
Longevity Without Cardiac Surgery

1. *Common defects* with expected adult survival but exceptional longevity.
2. *Common defects* with unexpected adult survival and exceptional longevity.
3. *Uncommon defects* with expected adult survival and exceptional longevity.
4. *Uncommon defects* with unexpected adult survival and exceptional longevity. The rarest of the rare.
Common Defects With Expected Adult Survival but Exceptional Longevity
Ostium Secundum ASD Age 85
Ostium Secundum ASD Age 95
Ostium Primum ASD Age 76
Common Atrium Age 57
Lutembacher Syndrome
Secundum ASD
Rheumatic Mitral Stenosis
Lutembacher is a German name, but Dr. Lutembacher used the French pronunciation (Loo-tem-bah-share) because he was Alsatian when Alsace-Lorraine was part of France. The pronunciation is now anglicized to Loo-tem-bah-ker.
Lutembacher’s original patient was a 61 year old woman who had been pregnant seven times. Firkett’s patient was a 74 year old woman who had endured 11 pregnancies. In one instance, an 81 year old woman with Lutembacher syndrome experienced no cardiac symptoms until her 75th year.
Iatrogenic Lutembacher Syndrome
Scimitar

Middle Eastern or South Asian sword with a curved blade
Scimitar Syndrome Age 63

Anomalous connection of the pulmonary veins in one lung to the inferior vena cava.
Pulmonary Valve Stenosis
Pulmonary Valve Stenosis

An appreciable number of patients with moderate to severe congenital pulmonary stenosis claim to be virtually symptom free. A group of patients with right ventricular systolic pressures of 75 to 100mm Hg included a New Zealand long-distance swimmer, a female athlete, an English hockey captain and a long-distance runner.

Paul Wood, OBE, MD, FRCP
The UCLA Registry includes a 17 year old boy with pulmonary valve stenosis who played baseball despite a right ventricular systolic pressure of 200 mm Hg, and a 32 year old man who had run the quarter mile in high school despite a resting right ventricular systolic pressure of 75 mm Hg.
Patent Ductus Arteriosus
Calcified Ductus Age 74
Calcified Patent Ductus Age 84
Coarctation Age 76
Complete Aortic Obstruction
Coarctation of the Aorta
U-Shaped Retinal Arterioles
Uncommon Defects with Expected Adult Survival But Exceptional Longevity
Anomalous Origin of LCA from PT
The Bland/White Garland Syndrome
Two Murmurs:
1) The Anomalous LCA
2) Mitral Regurgitation
A Distinctive Electrocardiogram
Left Axis Deviation
Left Ventricular Hypertrophy
**LCA from PT**

**Cultured Chick Cardiomyocytes**
Right Coronary Artery to Coronary Sinus Fistula
RCA to RA Fistula Age 74
SINUS of VALSALVA ANEURYSM

RA

Ao

PT

RV
Typical Sinus of Valsalva Aneurysm into Right Atrium

Age 32
Unusual Sinus of Valsalva Rupture into RV
Age 35
Asymptomatic Continuous Murmur
Rare Unruptured Sinus of Valsalva Aneurysm
Age 85
Pulmonary AV Fistula Age 73
Balloon Occlusion
Ebstein’s Anomaly
Ebstein’s Anomaly of the Tricuspid Valve

There are legendary accounts of astonishing longevity with survival into the eighth and ninth decades. Ebstein’s anomaly was discovered at necropsy in a 75 year old man who, in his youth, had been a lumberjack working on log booms. He was asymptomatic until his fifties, when he was obliged to outrun an irate female bear. At necropsy 25 years later, the tricuspid valve was malformed, and his right atrium was thin-walled and greatly dilated. The oldest recorded patient with Ebstein’s anomaly lived to age 85 years with no cardiac symptoms until age 79 when he developed right ventricular failure.
Ehstein’s Anomaly WPW Age 62
Ventricular Inversion

Ist der rechte Ventrikel der richtige Ventrikel?

Is the right ventricle the right ventricle?

Prof. Dr H. C. Kallfelz Hannover

The vulnerable chamber is the subaortic morphologic right ventricle which is perfused by a morphologic right coronary artery and has an inherently lower ejection fraction than a morphologic left ventricle.
Left-sided Ebstein’s Anomaly
Congenitally Corrected TGA
Left-Sided Ebstein’s Anomaly
Ventricular Inversion
Right to Left Septal Depolarization
Common Defects with Unexpected Adult Survival and Exceptional Longevity
Complete Transposition of the Great Arteries

Death Rates Without Intervention:
First week 30%
First month 50%
First year 90%
Complete Transposition of the Great Arteries Age 18 Months
Against All Odds
Age 36 Years
Common Defects with Unexpected Adult Survival and Exceptional Longevity
We have seen from our observations that cyanosis, especially in the adult, is the result of a small number of cardiac malformations well determined. One of these cardiac malformations is much more frequent than others.

Arthur Fallot 1888
### Fallot’s Tetralogy

#### Survival Without Intervention

<table>
<thead>
<tr>
<th>Age</th>
<th>Survival Rate</th>
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<tbody>
<tr>
<td>1st birthday</td>
<td>66%</td>
</tr>
<tr>
<td>Age 3 years</td>
<td>50%</td>
</tr>
<tr>
<td>Age 10</td>
<td>25%</td>
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<td>Age 20</td>
<td>11%</td>
</tr>
<tr>
<td>Age 30</td>
<td>6%</td>
</tr>
<tr>
<td>Age 40</td>
<td>3%</td>
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Fallot’s Tetralogy/Pulmonary Atresia
Age 56
Oldest Unoperated Tetralogy

Surgical Repair of Tetralogy of Fallot in a Seventy-five Year Old Patient,” the oldest known unoperated survivor and the oldest to undergo intracardiac repair. *International Journal of Cardiology* 2008.
“Increasing geriatric clinical and pathological experience is beginning to shed light on a host of problems relating to the life history of cardiovascular disease. In any large series of geriatric necropsies, for example, atrial septal defect is always well represented, but where’s the Maladie de Roger? Assuming it does provide immortality, it must either close spontaneously in middle life or have long since run its mortal course.”

Paul Wood, O.B.E., M.D., F.R.C.P.
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Necropsy VSD Age 68
Closed Inlet VSD
A Rarity
Moderately Restrictive VSD Age 46
Eisenmenger VSD Age 43
Eisenmenger
VSD
Age 54
Eisenmenger VSD Pulmonary Aneurysms
Age 54
Thrombosis in Dilated Hypertensive Proximal Pulmonary Arteries. A Therapeutic Dilemma
Aortopulmonary Window Age 57
Aortopulmonary Window
Age 61

A-P Window
The Rarest of the Rare

Uncommon Defects with Unexpected Survival and Exceptional Longevity
Truncus Arteriosus

Age 10

Age 42
Isolated Quadricuspid Aortic Valve

Age 56

Age 62
Eisenmenger Truncus
Intrapulmonary Hemorrhage Age 55
Truncus Arterosus
Age 54
Heterotaxy Left Isomerism
Left Isomerism
18 Months
Left Isomerism
Age 52
If as many as 90% of infants born with anomalous origin of the left coronary artery from the pulmonary trunk die in their first year, why did a patient of Maude Abbott’s live to age 60 years? Why does the same disease express itself so differently?
Unlocking genomic information has been considered the key to understanding cellular and molecular mechanisms, but it is now clear that knowledge of the DNA sequence is essential but insufficient. A more meaningful understanding of gene expression can be achieved through characterization of the protein products of that expression—-the ultimate biological determinants of disease phenotype. The term *proteome* was coined to describe the proteins encoded from a specific genome. The new discipline is called *proteomics.*
Longevity In Unoperated Congenital Heart Disease

Never Make Predictions, Especially About the Future
Mitral Stenosis
SALLE HENRI ROGER
Uncommon Defects with Expected Adult Survival But Exceptional Longevity
The natural history of any disease is a description of what happens to people with that disease who do not receive treatment for it.

Julien I. E. Hoffman
“Natural” History

- Pharmacologic therapeutics are not natural.
- Anticoagulants are not natural.
- Pacemakers are not natural.
- Electrical cardioversion is not natural.
- Electrical defibrillation is not natural.
- Radiofrequency ablation is not natural.
- So – naturally, natural history is inappropriate in contemporary medicine.
Categories

1. *Unoperated* – Unrecognized or inoperable. Longevity improved by medical management.

2. *Postoperative* – Improved longevity after operation is not due to operation alone.