**Tetralogy of Fallot Clinical Research Study**

- **Research Goal:** Test a new way to look at the right side of the heart by new ECHO technology and compare that with MRI.
- The study visit will last approximately 3 hours and involves a cardiac MRI and ECHO.
- 9 years old and up with Tetralogy of Fallot.
- No devices such as ICD or Pacemaker.
- No RV to PA conduit.
- For more information, contact T. Aaron West at thomas.west@nationwidechildrens.org or call (614)355-3448.
- Research incentive offered.
Tetralogy of Fallot
Past, Present, Future

Stephen R. Crumb, APN
Coordinator, COACH Program
Columbus Ohio Adult Congenital Heart
Disclosure: Go Buck(y)(y) (s)?
Survival to 18 years of age with CHD

<table>
<thead>
<tr>
<th>Year Born with CHD</th>
<th>1940</th>
<th>1960</th>
<th>1970</th>
<th>1980</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>20</td>
<td>40</td>
<td>75</td>
<td>90</td>
</tr>
</tbody>
</table>

Year Born with CHD


Survival rates:
- 1940: 20
- 1960: 40
- 1970: 75
- 1980: 90

Nationwide Children's

When your child needs a hospital, everything matters.
Pediatric to Adult Congenital Heart Disease

Expanding Population of Adolescents and Adults with CHD

Increased Mid Term Survival

Increased Early Survival

Lower Perioperative Mortality

Early Complete Repair

Improved Surgical Techniques

Advances in NICU Care

Fetal Diagnosis

Incidence of CHD

Nationwide Children's Hospital

When your child needs a hospital, everything matters.

Ohio State University
Ratio of Pediatric to Adult Patients with CHD

- Pediatric patients
- Adult patients

1965, 1985, 2005
Normal Heart

Tetralogy of Fallot

ETIENNE-LOUIS ARTHUR FALLOT

Mixing red and blue blood
Blue Baby Syndrome
A Success Story—Blalock-Taussig-Thomas and Blue Baby Syndrome

Alfred Blalock, MD  Vivien Thomas  Helen Taussig, MD
Something the Lord made
That First Operation

On **November 29, 1944**, Drs. Blalock and Taussig decided to proceed with the anastomosis, or joining, of the subclavian artery to the pulmonary artery in a cyanotic child.

Dr. Helen Taussig was convinced that the operation would help the patient, and despite the technical problems of operating on a very small and very ill child, Dr. Alfred Blalock's skill was equal to the task. Blalock worked with his surgical team and his invaluable assistant **Vivien T. Thomas**, who stood behind Blalock and offered a number of helpful suggestions in regard to the technique employed.

The tiny child who had been at such great risk survived the operation and slowly improved. Over the succeeding days she gradually became less blue. By the end of the second postoperative week it was clear she would recover.

*When I saw Eileen for the first time, it was like a miracle... I was beside myself with happiness.*

--mother of the first child to receive a Blalock-Taussig-Thomas shunt
The Operation

Vivien Thomas (top left) stands behind Dr. Alfred Blalock during an early operation at Johns Hopkins Hospital.

Courtesy Alan Mason Chesney Medical Archives, Johns Hopkins University
The Blalock-Taussig-Thomas Shunt

This procedure diverts blood from an aortic branch to the pulmonary artery, allowing blood to flow to the lungs to receive oxygen.
First Report

Journal of the American Medical Association
May 19, 1945

THE SURGICAL TREATMENT OF MALFORMATIONS OF THE HEART
IN WHICH THERE IS PULMONARY STENOSIS
OR PULMONARY ATRESIA

ALFRED BLALOCK, M.D.
AND
HELEN B. TAUSSIG, M.D.
BALTIMORE
### From Palliation to Repair

**Surgical Repairs in Tetralogy of Fallot**

<table>
<thead>
<tr>
<th>Surgical Procedure</th>
<th>Description of Surgery</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Pulmonary valvotomy</strong></td>
<td>Opening-up of a stenotic pulmonary valve</td>
<td>Augments blood flow to the lungs</td>
</tr>
<tr>
<td><strong>Infundibular resection</strong></td>
<td>Removal of muscular tissue below the pulmonary valve</td>
<td>Augments blood flow to the lungs</td>
</tr>
<tr>
<td><strong>RVOT patch</strong></td>
<td>Patch placed across the RVOT</td>
<td>Augments blood flow to the lungs</td>
</tr>
<tr>
<td><strong>Transannular patch</strong></td>
<td>Patch placed across the narrowed pulmonary valve</td>
<td>Disrupts the pulmonary valve; may result in severe pulmonary valve leaking</td>
</tr>
<tr>
<td><strong>Pulmonary valve replacement</strong></td>
<td>Human or porcine (pig) tissue valve is used to replace the original pulmonary valve</td>
<td>Used in adolescents and adults to treat pulmonary valve leaking (regurgitation)</td>
</tr>
<tr>
<td><strong>Extracardiac conduit</strong></td>
<td>Conduit (or tube) connecting the right ventricle to the pulmonary artery</td>
<td>Treatment of pulmonary atresia</td>
</tr>
<tr>
<td><strong>Angioplasty or patch augmentation</strong></td>
<td>Repair of pulmonary arteries</td>
<td>Treatment for hypoplastic (small) main pulmonary artery or stenosis of central pulmonary arteries</td>
</tr>
</tbody>
</table>
Direct Vision Intracardiac Surgical Correction of the Tetralogy of Fallot, Pentalogy of Fallot, and Pulmonary Atresia Defects
Report of First Ten Cases

C. WALTON LILLEHEI, M.D., MORLEY COHEN, M.D., HEBERT E. WARDEN, M.D., RAYMOND C. READ, M.D., JOSEPH B. AUST, M.D., RICHARD A. DEWALL, M.D., AND RICHARD L. VARCO, M.D.

Minneapolis, Minn.
From the Department of Surgery, University of Minnesota Medical School, Minneapolis
Cross-Circulation as the first heart-lung machine
The age of mechanical heart-lung bypass
Repair of tetralogy of Fallot

Tetralogy of Fallot (TOF) Interior View

- Atrial Septal Defect (ASD)
- Tricuspid Valve
- Small Pulmonary Valve
- Narrowed Pathway from RV to PA (Right Ventricular Outflow Tract - RVOT)
- Opening Between Ventricles (Ventricular Septal Defect - VSD)
- Mitral Valve
- Aortic Valve

Repair of Tetralogy of Fallot (TOF) Exterior View

- Patch to Enlarge Narrowed Pathway from RV to PA
- VSD Patch (To Close Opening Between Ventricles)
Repair is still not cure!

Leaking of the pulmonary valve
Enlargement of the right ventricle
Weakening of the right ventricle
Right sided heart failure
Narrowing of the lung arteries
Need for multiple operations during lifetime
Arrhythmias
Sudden death
Right ventricular enlargement
Treatment of left-over heart problems
Stent therapy of a narrowed pulmonary artery
What does the human PV look like?
Surgical Pulmonary valve replacement

Pulmonary Homograft

Hancock valved conduit

Pericardial valve
Transcatheter pulmonary valve replacement

Melody Valve
- Bovine jugular venous valve segment
- Platinum-Iridium stent

Image 4a
Melody valve inside tissue valve

Tissue valve narrowed
Melody Valve placement
The Melody pulmonary valve
Limitations of the Melody Transcatheter Pulmonary Valve

The Melody TPV only can treat 15% of the patients that need it (only 15% have RV – PA Conduit)

What about the 85% of patients with native RVOT reconstruction ???

• 1st in Man…Philip Bonhoeffer, GOSH, London

• Native Outflow Tract TPV Clinical Research Centers were recently selected
  • NCH/OSU was chosen as the USA site, while Toronto Sick Kids chosen for Canada, and Munich chosen for Europe…to be started in 1st quarter 2012
First in Man: Philip Bonhoeffer
Native right ventricular outflow tract stent / valve
- Adapts to irregularities of patient anatomy
- Increases access to non-surgical care for many patients.
Enough technology—what about living with tetralogy of Fallot?

- At least 90% survival 30 years after operation
- Studies indicate that academic achievement is average or above
- Generally satisfied with life
- May be somewhat underemployed
- Severity of congenital heart disease is not associated with lower quality of life
- Pregnancy can be safe and successful with appropriate care
Living Long and Well with tetralogy of Fallot

Regular follow-up with ACHD team

- Routine physical exam, ECG, oximetry
- Echocardiogram to monitor heart function and blood flow through pulmonary valve
- Periodic Holter monitor to look for arrhythmias
- We may recommend MRI or CT Scans to get more detailed information about your heart
Living Long and Well with tetralogy of Fallot

- You may need cardiac catheterization to evaluate and treat:
  - Narrowing in the pulmonary artery
  - Arrhythmias (Ablation)

- You may need to have more surgery
  - Pacemaker / ICD placement
  - Valve replacement
Living Long and Well with tetralogy of Fallot

- Listen to your body
  - Call us with any symptoms or concerns
- Take your medications as prescribed
- Avoid tobacco products
- Get regular exercise
- Eat healthy
  - Fruits and vegetables
  - Whole grains
  - Go easy on fats, processed carbohydrates and salt
What does the future hold for people with tetralogy of Fallot?
Final Thought

May you live long and well enough to visit the “Golden Years Clinic on Century Square for Spleen Readjustment and Muffler Repair”
Team COACH