From the desk of Hon. Editor:

“The heart (is) deceitful above all (things), and desperately wicked: who can know it?”

- Jeremiah 17:9

Dear Friends,

10 out of 1,000 children born have Congenital Heart Disease (CHD). Most of them require treatment, either by catheter intervention or surgery, some even in neonatal period & majority during infancy or childhood.

With the help of a team approach in presence of sound infrastructure, we have been able to reach international standards in the care of these delicate and often fragile babies.

In this issue of Healthy Heart, we discuss Tetralogy repair in infancy, analyse a rare case of pulmonary artery sling treated successfully & peep into the fascinating & adventurous history of modern pediatric cardiac surgery.

- Dr. Shaunak Shah

Tetralogy Repair in Infancy

Tetralogy of Fallot (TOF) is the commonest cyanotic congenital heart disease.

Briefly, the anterior and cephalad displacement of conal septum results in varying degree of right ventricular outflow tract (RVOT) obstruction. Hypoplasia of pulmonary annulus and branch pulmonary arteries is common. A large ventricular septal defect with RVOT obstruction results in Right to Left shunt with varying degree of cyanosis.

Babies born with TOF may present with cyanotic spells either in neonatal age or infancy. Cold weather, dehydration aggravate the infundibular spasm.

When an infant presents with spells, there is often therapeutic dilemma.

In the past, most patients were treated with palliative Blalock-Taussig-Thomas shunt in infancy and were offered complete correction at around 3 or 4 years of age. The problem with shunt is: technical difficulties, distortion of branch pulmonary arteries, volume overload,
and development of chest wall collaterals which make subsequent surgery difficult.

Early correction in infancy on the other hand has the advantage of a physiologically & anatomically normal heart with normal growth in children & no need for the second operation.

Only a few years back, it was believed that Tetralogy correction in infancy is difficult (if not impossible). Even in the West, only few centers would consider doing tetralogy correction in infancy, because of high mortality and morbidity.

Following is a list of steps that we have taken to make TOF repair in infancy a possibility:

1) Accurate pre-op diagnosis: With the help of a detailed 2D Echo examination, supplemented with CT pulmonary angiography in some cases, an accurate anatomy is defined. This means that virtually no kid requires cardiac catheterization & angiography. Cath+angio is needed in patient who has significant collaterals which are embolized just prior to surgery.

2) New anesthesia protocols: Avoidance of spells during induction, newer & safer anesthetic agents, use of blood preservation techniques.

3) Technical improvements: This includes gentle handling of tissues, interrupted sutures for VSD closure, respect of tricuspid valve, avoidance of trans annular patch, & limited trans annular patch, wherever necessary.

4) Advances in extracorporeal technology: like use of arterial filter, del Nido cardioplegia, hemofiltration, avoidance of hypothermia.

5) Intra operative Echocardiography either Trans esophageal or Epicardial at the end of surgery: This ensures that no patient leaves operation room with a residual defect.

6) Aggressive ICU Care: This includes judicious use of blood & blood products, early extubation, use of inodilators like milrinone, & peritoneal dialysis.

With these policies, we no longer offer palliation (BT shunt surgery) to children with TOF in infancy except in the following condition:

1. Weight below 4 kg & age below 4 months.
2. LAD (Left Anterior Descending Artery) crossing RVOT with hypoplastic annulus.
3. Multiple VSDs.
4. Branch PA anomalies.

Out of the 33 TOF repair that we did in 2014, seven were infants weighing from 5.5 to 7 kg. There was no death in this group and one patient had prolonged ICU Stay.

As our experience and expertise improve, we should be able to offer TOF correction, even to neonates in near future.
Thirty-one days old, 3kg child was brought to CIMS from other NICU (Neonatal Intensive Care Unit) with respiratory distress and pneumonia X-ray chest showed left midzone consolidation. A CT scan (Figure-1) showed Left Pulmonary Artery (LPA) sling in which LPA, instead of arising as a branch of main pulmonary artery, came from mid Right Pulmonary Artery (RPA), circled around trachea, (between trachea & esophagus) & reached left lung hilum. Because of anomalous course, LPA pressed on trachea & child developed respiratory distress.

A 2D echo suggested secundum ASD & patent ductus arteriosus in addition to LPA sling. A fibre optic bronchoscopy (FOB) showed pulsatile compression close to carina.

The child underwent surgical correction: through mid sternotomy, cardiopulmonary bypass was initiated, PDA was divided, LPA was divided at its origin from RPA, mobilized from behind & brought in front of left main bronchus and reanastomosed to MPA using fine sutures in posterior continuous & anterior interrupted fashion. The ASD was closed directly. The child made an uneventful recovery and was discharged from hospital on the 12th post operative day.

On follow-up, child is symptom free & has put on 700 gms weight over a period of 1 month.
On 24th August, 1938, Robert Edward Gross ligated a Patent Ductus Arteriosus (PDA) in a four year old girl & began the era of modern cardiac surgery. Gross was senior resident in Boston Children Hospital at that time and did this operation when his chief, William Ladd was away on vacation. This news rattled Helen Taussig, a Pediatric Cardiologist.

Taussig had observed in her studies that children with Tetralogy of Fallot survived longer if they had a PDA. She wondered if a duct can be closed, surely a new one can be created.

She went to meet Dr. Gross in Boston. However success had gone to his head who retorted: “I close ductuses, I do not make them”.

Disappointed, Taussig went to Alfred Blalock in Baltimore who was working at that time on systemic to pulmonary artery connection. Blalock liked the idea and working with his assistant, Vivien Thomas did first successful shunt operation at Johns Hopkins Hospital. The success of this operation resulted in a stream of blue babies coming to Baltimore from all over world.

Blauflock and Taussig became famous but Thomas' contribution was overlooked.

African – American Thomas was a Laboratory Technician, all the technical details were the result of his hard work in Animal Lab. He was present during first Shunt operation and had guided Blalock throughout the surgery.

Later, however his role was appreciated & in 2004 HBO made a film : “Something the Lord Made”. Johns Hopkins even gave him a honorary doctorate & his statue has been put up in the hospital campus.

“An idea whose time has come is never wasted, If you don’t grab it, someone else will”
What is heart failure?
Heart failure is a widespread, chronic condition that develops when the heart muscle weakens and is unable to pump a sufficient amount of blood throughout the body. Heart failure worsens over time and is typically caused by persistent high blood pressure, heart attack, valve disease and other forms of heart disease or birth defects. Left untreated, the lack of adequate blood flow causes the organs to progressively fail, resulting in numerous medical complications that deteriorate a person's quality of life and often leads to death.

If you or someone you know has heart failure, also called congestive heart failure, you are not alone. According to the American Heart Association, more than five million Americans are living with heart failure, with about 670,000 new cases diagnosed every year.

People with heart failure often have shortness of breath and fatigue. Years of living with blocked arteries or high blood pressure can leave your heart too weak to pump enough blood to your body. As symptoms worsen, advanced heart failure develops.

Advanced heart failure is a serious condition. Though you may find ways to relieve worsening symptoms, such as being less active, drinking less liquid, and sleeping with pillows, your daily enjoyment of life can suffer. The key to managing advanced heart failure is to take control. It’s up to you to follow your physician’s recommendations and make the necessary lifestyle improvements for healthy living.

About Ventricular Assist Device (VAD) Therapy
Restoring Hope with Mechanical Circulatory Support

Mechanical Circulatory Support (MCS) uses blood pumps called VADs to improve blood flow. It assists the heart to pump blood to the body. It does not replace the heart. Patients must have surgery to implant the device. For patients waiting for a heart transplant, a VAD may help them survive until a donor heart is found. This is known as Bridge-to-Transplantation. Some advanced heart failure patients may not be candidates for a transplant because of other diseases or age. The patients may benefit from long-term VAD support. This is called Destination Therapy. Occasionally, patients’ hearts get better with a VAD. That’s because the pump gives their heart a chance to “rest.”

What is an LVAD?
LVAD stands for Left Ventricular Assist Device. It is a mechanical device that circulates blood throughout the body when the heart is too weak to pump blood on its own. It is sometimes called a “heart pump” or “VAD.” HeartMate II is a miniaturized implantable LVAD that represents a breakthrough in medical technology and has rapidly become the most widely used device of its kind in the world.

How does HeartMate II work?
HeartMate II is designed to take over the pumping function of the patient’s left ventricle. The device is placed just below the diaphragm in the abdomen. It is attached to the left ventricle, and the aorta, the main artery that carries oxygenated blood from the left ventricle to the entire body. An external, wearable system that includes a small controller and two batteries is
attached by an external driveline. The wearable system is either worn under or on top of clothing.

How does HeartMate II help a heart failure patient?
HeartMate II is designed to restore blood flow throughout the body, enabling the patient to breathe more easily and feel less fatigued. The patient’s organs will receive more blood than they did before receiving the LVAD, and this will likely improve their organ function. After receiving an LVAD, patients generally feel more energetic and are able to resume normal activities that they were unable to do prior to receiving the device.

How active can patients be with the HeartMate II?
Because patients are in a severe stage of heart failure before receiving the device, they are very debilitated and typically very limited in terms of activity level. After receiving HeartMate II, the majority of patients can return to their favorite daily activities, with the primary limitation being water immersion. Many patients are able to return to work and resume hobbies that they haven’t been able to do for years.

Is HeartMate II a good treatment option for advanced heart failure patients?
Yes. HeartMate II is considered a standard of care in cases of advanced heart failure. Studies have shown that advanced heart failure patients treated with an LVAD can live longer and enjoy a much improved quality of life compared with those being treated with drug therapy alone. There are approximately 50,000 – 100,000 advanced heart failure patients who could benefit from an LVAD in the U.S.

How long do the batteries last?
The latest generation of batteries used to support LVADs may last up to 14 hours before needing to be recharged.

Who can get a HeartMate II?
Patients suffering from advanced heart failure and who have exhausted the limitations of medical therapies may be candidates to receive a HeartMate II. Due to the device's ability to allow the patient's heart to rest and take over the pumping function, it has been shown that the LVAD provides the opportunity for a weakened heart to regain some of its own function. Patients should consult a physician to find out if they are a candidate for LVAD therapy.

Does someone with a HeartMate II still have a pulse?
A patient who is implanted with a HeartMate II usually has a dampened pulse. The reason for this is that HeartMate II moves blood from the heart to the body continuously along with the heartbeat. The “strength” of the patient’s pulse will depend on how much assistance the LVAD is providing to the heart.

How big are LVADs?
LVADs vary in size, but the HeartMate II - the smallest of all the FDA-approved LVADs – measures approximately 3 inches in length and weighs approximately 10 ounces.

Where can I get an LVAD?
There are more than 200 centers worldwide that are implanting HeartMate II. Now CIMS hospital is certified for implantation.

Dr. Dhiren Shah, Dr. Milan Chag & Dr. Niren Bhavsar are qualified & certified for this procedure

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The award selection process was conducted by a panel of independent judges and the winners were selected through a process based upon a combination of surveys and jury selection (across 150 best hospitals in India and 10 in Ahmedabad), adjudicated by a research & survey process carried out in 4 phases in the doctor patient community by Times Group, Ernst & Young and supported by Nielsen group.
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