HEALTHY HEART

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As over 85% of children undergoing Congenital Heart Disease (CHD) surgery or intervention survive into adulthood, we are witnessing a large number of adults with CHD. Also, in India we are increasingly witnessing elderly patients diagnosed with CHD for the first time. This is a group of patients who present with unique cardiac, non cardiac & social problems.

In this issue, we discuss how to tackle these problems & a need for a Subspecialty of Adult Congenital Heart Diseases (ACHD) or Grown Up Congenital Heart Disease (GUCH).

ADULT CONGENITAL HEART DISEASE (ACHD)

Figure : 1

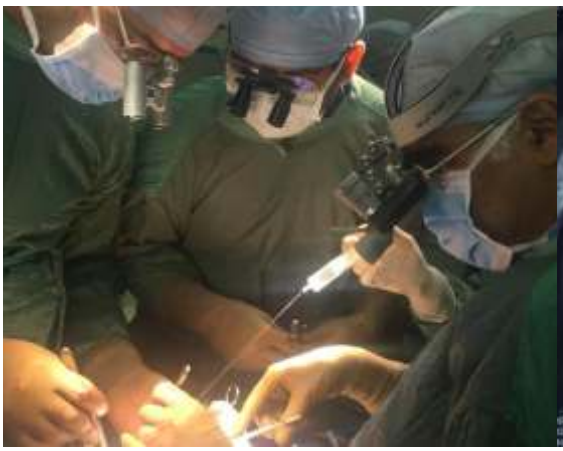
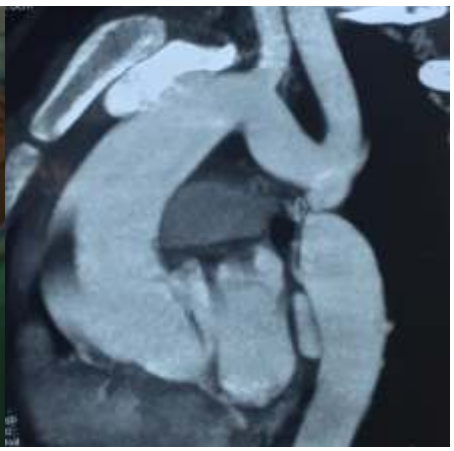


Figure : 2 Adult Coarctation



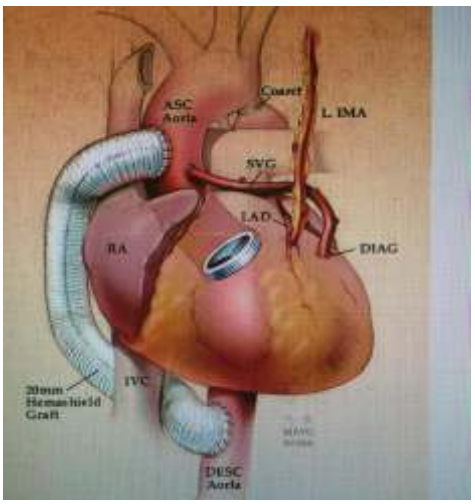


Figure : 3 Extra anatomic repair of Adult Coarctation

Case 1 : Twenty eight year old Amita, developed fever during the 5th month of her first pregnancy. An alert physician noticed murmur in the precordium & a 2 D Echo was done. She was diagnosed to have Ventricular Septal Defect (VSD) with Aortic Regurgitation (AR). There was a suspicion of vegetation. Because of progressive heart failure & pyrexia, a decision was made to operate upon her. With continuous double fetal monitoring (she had twin pregnancy) &

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under cardiopulmonary bypass, she underwent Open Heart Surgery. There was Ruptured Sinus of Valsalva (RSOV) and vegetation on pulmonary valve. She underwent RSOV repair + VSD Closure + Removal of Vegetation. She had an uneventful recovery & was discharged from the hospital on the eighth day. She delivered two normal babies at term.

Case 2: Sonia was seven years old in 2000, when she had open heart surgery with Tetralogy of Fallot (TOF) repair. In 2019, she delivered a healthy child at full term.

Case 3: Mrs. Shyama, 61 year old, developed breathlessness on mild exertion while on a trip to a hill station. Investigations revealed her to have a large sinus Venous Atrial Septal Defect (VASD) with moderate pulmonary hypertension. She had open heart surgery with Warden procedure.

So, what is Adult Congenital Heart Disease (ACHD) or Grown Up Congenital Heart Disease (GUCH) ?

Congenital Heart Disease in the adult is the presence of unrepaired or repaired congenital heart disease in patients aged 21 years or older.

Extent of the problem:

The incidence of congenital heart disease in society is 8 to 10 per 1000, some of which may require surgery. Since the results of surgery for CHD are improving, a large number of patients

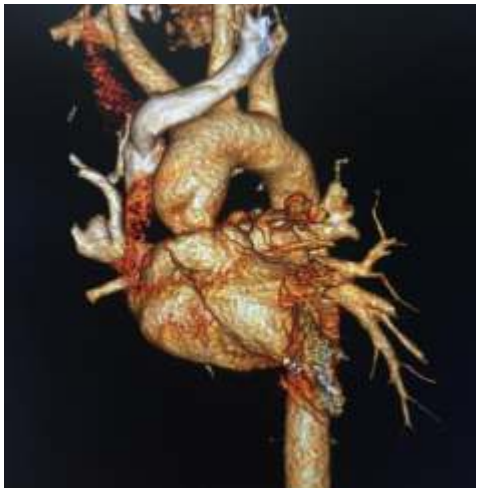

Figure : 4 Supravalvar Aortic Stenosis in a 27 years old woman. She had Brom's 3 patch repair.

who have undergone surgery in childhood are likely to grow into adults. Except for uncomplicated Patient Ductus Arteriosus (PDA) & Atrial Septal Defect (ASD), most cardiac surgeries are considered palliative rather than curative. As a result, we are likely to encounter large population of patients having corrected CHD, requiring medical follow up.

Also, many CHDs remain asymptomatic / mildly symptomatic & patient present first in adulthood. Common CHDs presenting in late adulthood are : Atrial Septal Defect (including sinus venous ASDs), VSD with PS, Adult Tetralogy, Ebstein's Anomaly etc,.

A third group of patients knew about CHDs in pediatric age group, but did not under go repair for some reasons.

Problems with ACHD

In corrected CHD in childhood, the adults may present with (A) Residual defect (B) New defect or (C) Social / occupational problem.

(A) Residual Defect:

Like small VSD, Right Ventricular Outflow Tract Obstruction (RVOTO), Progressive Aortic Regurgitation (AR). Some of this may need re-intervention.

(B) New Lesion:

Some examples of this problem are: progressive Pulmonary Regurgitation (PR) leading to Right Ventricular (RV) dysfunction in an operated case of TOF., AR in dilated aorta in Tetralogy, calcification & narrowing or leak in conduit & its valve.

(C) Rhythm disturbances :

Some patients develop complete heart block either as a part of natural history of disease: (e.g. corrected Transposition of great arteries) or after intervention for CHD (post device closure or open heart surgery for VSD closure, SAM resection, TOF repair). Many of these patients will need a regular change of pacemaker. Also, large left to right shunt patients may have atrial arrhythmias because of large size of atria.

(D) Progressive Heart Failure :

Uncorrected CHD, an incomplete correction with residual defect, or surgery in previous era may predispose to progressive worsening of myocardial dysfunction: especially a long ventriculotomy for TOF repair or residual valve leak or stenosis.

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(E) Pulmonary hypertension & Eisenmenger syndrome in uncorrected large L to R lesions like VSD, ASD, PDA

(F) Surgical Issues:

- Collaterals in cases with reduced pulmonary blood flow: It is important to identify these collaterals before surgery so that they can be either embolized or taken care off during surgery.
- Re-do sternotomy: A Previous surgery produces adhesions around heart. A careful dissection, femorofemoral bypass are few of the tricks to circumvent this.
- Repeated cardiac catheterization may have damaged femoral vessels & it is essential to get a Doppler study of femoral vessels prior to surgery.
- Some patients are on antiplatelet or anticoagulation which need to be discontinued prior to surgery or procedure. Or else they will require multiple transfusions or procoagulants.

(G) Non-Cardiac issues like pregnancy, contraception & dental procedures.

(H) Social Problem:

- Marriage in a patient operated for CHD in childhood
- Exercise / sports activity in patients who have undergone CHD repair
- Occupation choices

So who should look after patient with ACHD ?

Obviously, a team of Pediatric cardiologist, cardiac surgeon trained in pediatric cardiac surgery, electrophysiologist, and internist, should be taking care of patients in A Specialty ACHD clinics.

In the end, lets us discuses some interesting scenarios :

- A 23 year old employee is found to have Congenital Bicuspid Aortic Valve (CBAV) in his pre-employment check-up without Aortic Stenosis.
  (A) Should his employer be worried?
  (B) Can he be given a work which involves strenuous manual labour ?
  (C) What advise will you give to the employee ?

- Twenty one year old Akshay wants to join armed forces. He had undergone Atrial Septal Defect repair when he was 3 year old is he fit to join armed forces ?

- Twenty five year old Barkha had undergone Tetralogy repair in childhood. Now she is in first trimester of pregnancy. What additional precautions should she take ? What extra tests would you advice her ?

- 22 Year old Sujata had Fontan Operation for Tricuspid Atresia type 1 B when she was 13 year old. Her parents are looking for a suitable boy for her. The prospective in laws decide to meet you & ask : Sir, is this marriage possible ?

- A young cadet dies after exercise in a parade ground. You are asked to give a death certificate. Will you oblige ? What is the cause of death ?

- A newly wed women wants contraception advice. She had undergone Tetralogy repair when she was an 8 year old, what is the method of choice for her ?

Clearly, ACHD is a subspecialty which require a multi disciplinary team approach, for better quality of life for patients.

Considered by many to be the most beautiful film star, Madhubala (1933-1969), had an uncorrected Ventricular Septal Defect (VSD) & died of Eisenmerger's at 36 years of age.

Figure : 5

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- Chest pain or pressure
- Tightness, burning or heaviness in the chest
- Shortness of breath
- Profuse sweating
- Nausea and vomiting

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A Living Donor Liver Transplant on a 2 year old child
Youngest to receive this treatment in Gujarat
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Ahmedabad Mirror

'Listen to them and understand them'

Gujarat registers its youngest organ recipient

Two 2-year-olds give and receive life via two organ transplant surgeries

સીમ્સ હોસ્પિટલમાં 12 કલાકે સફળ ટ્રાન્સપ્લાન્ટ

2 વર્ષની બાળકીને માતાએ લિવરનો હિસ્સો દાન કર્યો

દંપતી આ બીમારીથી અગાઉ બાળક ગુમાવી ચૂક્યું હતું

સિમ્સ હોસ્પિટલના ગેસ્ટર, પેડિયાટ્રિકલના 30 સભ્યની ટીમે 2 વર્ષની બાળકી પર 12 કલાકની સફળ સર્જરીને અંતે લિવર ટ્રાન્સપ્લાન્ટ કર્યું છે. ટ્રાન્સપ્લાન્ટ પહેલાં ગેસ્ટરો-સ્ટાફના કોરોના ટેસ્ટ કરાયા હતા. બાળકી અને માતાનું બ્લડગ્રુપ અલગ હોવાથી 'પ્લાઝમા ફેરાસીસ'થી ટ્રાન્સપ્લાન્ટ પહેલાં ડાયાલીસીસથી બાળકીનાં લોહીમાંથી એન્ટિબોડી દૂર કરાયા હતા. ટ્રાન્સપ્લાન્ટમાં સિમ્સ ફાઉન્ડેશન, મિલાપે આર્થિક સહાય કરી હતી.

સિમ્સ હોસ્પિટલના લિવર ટ્રાન્સપ્લાન્ટ સર્જન ડો. આનંદ ખખ્ખરે જણાવ્યું હતું કે, બાળકી હિરવા લિવરની વારસાગત બીમારી 'સિટ્રુલિનેમિયા ટાઈપ-1'થી પીડાતી હતી. અગાઉ આ બીમારીથી એક બાળક ગુમાવી ચૂક્યા હતા.

સિમ્સના ચેરમેન ડો. કેયૂર પરીખ જણાવ્યું કે, બાળકીની માતાએ લિવરનો હિસ્સાનું દાન કર્યું, અને ગેસ્ટરો-સ્ટાફની ટીમે અશક્ય લાગતું ટ્રાન્સપ્લાન્ટ પાર પાડ્યું છે. બાળકીના પિતાના આર્થિક સ્થિતિને ધ્યાનમાં રાખી સિમ્સ ફાઉન્ડેશન, મિલાપે આર્થિક સહાય કરી છે.

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