A General IntroductionCommon operations performed in Paediatric Cardiac Surgery and Follow up Guidelines

1. Closed Heart Surgery: Surgery on Heart or Great Vessels done without using the Heart – Lung machine.
2. **Open Heart Surgery:** Surgery done on the Heart or Great vessels using a Heart – Lung machine.

Salient points on certain common types of Congenital Heart Surgery:

Shunts: Blalock Shunts are connections between systemic arteries such as a subclavian artery with its respective pulmonary artery. Currently all such shunts are performed using a PTFE conduit. These are performed via a thoracotomy usually, though they could be performed via a sternotomy in certain indications.

Glenn Shunts are those where the SVC is connected to the respective pulmonary artery to increase pulmonary blood flow without causing volume overload of the ventricle like the Blalock shunts. This is usually an open heart procedure especially in young children though it could be attempted as a closed procedure in older ones. Performed through a sternotomy or a Right thoracotomy.

Coactation of aorta repairs: Depending on the anatomy the repairs performed are:

1. Resection of the coarctation segment and end to end anastamosis
2. Subclavian flap aortoplasty where the left subclavian artery is used as a flap to enlarge the coarcted segment
3. Patch aortoplasty: A diamond shaped patch of PTFE is used to enlarge the coarcted segment.
4. Interposition graft.
5. Bypass with a graft.

The latter two procedures are not usually done in children but is reserved for repairs in adults.

ASD Closures: Approached by:

1. Median Sternotomy
2. Right Antero-lateral Thoracotomy – as in young female patients giving them a sub-mammary scar.
3. Right posterolateralTheracotomy – in prepubertal female children as the scar is more cosmetic and does not interfere with breast development.

Small ASD's can be closed directly. Others will need a pericardial patch. Primum ASD and Sinus Venosus ASD are always closed with a patch.

VSD Closures: Is done with a prosthetic material like Dacron or PTFE patch (Gore – Tex). Can be approached by many routes depending on location. Most are approached through the Right atrium and tricuspid valve. Subarterial VSD's are approached via the pulmonary artery or by a tranverseventriculotomy. LV approach for multiple VSD's is not preferred nowadays due to LV dysfunction being seen post

op. Rarely VSD's are approached via the aorta as in repair of RSOV with VSD .
Heart block after VSD closure is seen in less than 1% of patients in today's practice. However, an inlet VSD and VSD's extending towards the crux of the heart are more prone to develop complete heart block after surgery.

Tetralogy of Fallot Repair: This involves RV outflow resection, closure of the VSD and enlargement of the RV outflow tract. The repair can be done via the Right atrium or the Right Ventricle. Depending on the adequacy of the pulmonary annulus and Main pulmonary artery a pericardial patch is either put across the RV outflow or across the RVOT and pulmonary annulus to enlarge it. In the latter case there will be pulmonary regurgitation post operatively which is well tolerated for a long time.

Transposition of great Arteries: (TGA) The problem here is thet the ventriculoarterial connections heve been reversed with the RV giving rise to the aorta and the LV giving rise to the PA. This could be associated with a VSD, PDA Pulmonary stenosis or Coarctation/ Aortic arch interruptions.
Timing of surgical corrections depend absence or presence of a VSD or a large PDA. The LV will regress in its ability to support a systemic afterload once the PVR starts to come down in the perinatal period. This will be delayed in the presence of a large VSD or PDA as these lesions will continue to subject the LV to a systemic pressure and retain its ability to handle the systemic afterload. Corrective surgeries are usually planned in the neonatal/ infant age groups as the rate of attrition and development of PAH is very early in these children.

Arterial Switch Operation: Switching the Great arteries to restore the normal Ventriculoarterial connections. The coronary arteries also need to be transferred in the process which is the most delicate part of the surgery . This operation is also called anatomical correction as the normal anatomy is restored. Long term results are the best with this form of surgery. In cases where there is no largeVSD or PDAthis operation is best done at birth or within 2 to 3 weeks of age at the most.

Atrial Switch Operation: (Senning or Mustard procedures) By means of a baffle the systemic v enous blood could be diverted to the LV across the mitral valve and the pulmonary venous blood into the RV across the tricuspid valve, thus 'physiologically correcting the circulation' as the venous blood goes to the lungs and the oxygenated blood to the aorta. These operations are considered today if the child presents beyond the age for arterial switch operation and the LV has regressed making it unable to support the systemic afterload. Long term results are beset with problems of atrial arrythmias and progressive deterioration of RV function.

Rastelli Operation: An operation which involves closure of a large VSD and establishing RV to PA connection with an external conduit, valved or non valved. Classically done for cases of TGA with VSD and PS, or VSD with Pulmonary atresia. Long term results and need for reoperation depend on how quickly the Conduit degenerates/calcifies or the child outgrowing it in size. Hence if the child is not very cyanotic operation is best delayed or temporized with a Blalock Shunt till the child is big enough to accept a reasonable sized conduit.

REV operation: Very similar to the Rastelli Operation, but where the VSD is closed routing the blood from LV to aorta and RV to PA connection being established **without** a conduit. Pulmonary regurgitation remains as a residual lesion. Long term results again depend on how the heart compensates for the PR. Typically done for cases where there is no suspition of Pulmonary hypertension such as TGA/VSD with PS, VSD with Pulmonary atresia, etc. The anatomy is another determinant forthis operation to be performed.

Fontan Operation: The Fontan principle involves redirecting the whole systemic return into the lungs bypassing the 'RV' and using the single ventricle present to support the systemic circulation. This results in non – pulsatile flow to the lungs and the whole circulation depends on the single ventricle functioning at a low enddiatolic pressure. The current procedures performed incorporating this principle are Total Cavopulmonary connection, &Extracardiac Fontan corrections using an extracardiac conduit. These operations can be done in a single stage or in two stages depending on the pulmonary artery sizes or anatomy, the first stage being a bi-directional Glenn shunt.

Conditions for which the Fontan operation is considered are Tricuspid atresia, Single Ventricles, Double Inlet Ventricles, or very complex cyanotic lesions with VSD's not being able to be routed to the aorta. For a good post Fontan result, the prerequisites are 1. Large pulmonary arteries 2. Low pulmonary arterial pressures 3. Good ventricular function 4. Competent avtrioventricular and aortic valves 5. No LV outflow obstructions.

This operation is essentially a palliative operation as it is done to relieve cyanosis and reduce the volume overload on the single ventricle. However in the long term, problems of atrial arrythmias, and protein losing enteropathies develop in most patientsleading to reduction in longevity.

Ross Operation: This is an operation done in children where an aortic valve replacement is needed as an isolated procedure or with correction of an associated anomaly. It consists of harvesting the native pulmonary valve and implanting it in the aortic position and replacing a homograft valve in the pulmonary position. Hence it is essentially a 'Pulmonary Valve Switch'. The advantage is that the native pulmonary valve grows with the child and in future as the child outgrows the homograft, replacing a right sided valve such as a homograft in this case is technically easier than a left sided one. It also avoids prosthetic valve replacement and the need for anticoagulation with its attendant complications. Though the surgery is technically more demanding with a higher risk, the long term result is expected to be better.

9. Interventions for Congenital Heart Disease:

The ability to dilate blood vessels percutaneously was first demonstrated by Dr Andreas Gruentzig in 1978. In 1982 Kanfirst demonstrated that a congenitally stenotic valve could be dilated using a cylindrical balloon. This opened up new

vistas in the trans-catheter treatment of congenital heart lesions. Rapid progress has been now made in 1) Our understanding of mechanism of success or failure of balloon dilation 2) Development of newer hardware like low profile balloon, extra support wires which has contributed to increase in success rate of the procedure 3) Understanding the unnatural history of various congenital heart lesions from the vast data bank of intervention registries of balloon dilation. As a consequence balloon dilation has become the first procedure of choice in the treatment of valvar pulmonary stenosis, and post surgical recurrent coarctation. Although an essentially palliative procedure for the treatment of valvar aortic stenosis and native coarctation beyond the neonatal period, non-randomized studies have shown results achieved that are similar to that of surgical correction. Balloon dilation of pulmonary artery stenosis, pulmonary vein, venous baffles have shown poor overall results. Balloon dilation for sub-pulmonary stenosis like tetralogy of Fallot is controversial.

The development of stents to keep dilated vessel open has also found its place in treatment of congenital heart diseases. Thus pulmonary artery stenosis, recurrent coarctation in adults, venous baffles obstruction, conduit stenosis, which responds poorly to balloon dilation alone, have shown promising early and mid-term results. The use of stents to keep the ductus open, recurrent coarctation in children, in right ventricular outflow tract obstruction is controversial.

Development of devices to achieve closure of cardiac defects is relatively new. Patent arterial duct was the first defect to be closed with the help of devices and has shown good results on long term follow up. Recently stainless steel coils have been used to close small ducts (up to 3mm) with very promising medium term results. Its use has also been extended to larger ducts (up to 7mm). Devices used for closing atrial septal defects, ventricular septal defects are undergoing rapid development. In atrial septal defects its use is restricted to defects in the fossa ovalis area with well-defined rims and in ventricular defects, to only those that are in the muscular septum. Most of the devices in use today for atrial septal closure are relatively new and await long-term results. The results achieved by non surgical treatment for many congenital heart lesions are essentially palliative and may require repeat interventions at some point during follow-up which increase the cost of treatment substantially. The reuse of balloons, guide wires in developing countries like ours help reduce the overall cost of the procedure.

1. Follow up after Congenital Heart Operations and Interventions:

Aftercare of the children who have undergone cardiac procedures and surgeries is as important, and in some occasions, more important than the actual procedure performed. The follow up of these children depends on the following:

1. The nature of the surgery performed (palliative or definitive repair). Some children would have undergone a palliative procedure as a first step towards a more definitive surgery to be done at a later date.
2. Post Op complications: Complete heart block (CHB), diaphragmatic palsy, chylothorax
3. The presence of residual cardiac defects following a repair. It could even be a small residual ventricular septal defect (VSD) that would be hemodynamically insignificant but would deserve infective endocarditis (IE) prophylaxis.
4. The “natural history” of the cardiac condition following the surgery. For example, the incidence of arrhythmias increases with age following Atrial switch (Senning’s) operation for transposition of great arteries.

The primary care pediatrician should be reasonably well versed with the cardiac anatomy and the ongoing physiology following the cardiac surgery. Some general rules applicable to all children following cardiac intervention/ surgery:

1. At the first contact following the cardiac procedure or surgery, check for the lower limb pulses as femoral vessel cannulation is invariably done both during catheterization procedures or cardiac surgery.
2. Persistence of cardiac murmur is common despite complete repair and one should make a note of it. Development of a new murmur or worsening of the preexisting murmur warrants thorough re-evaluation by the cardiologist.
3. All common ailments should be treated as it is done for any other child. Even after complete repair of the cardiac defect, children may develop as many as 3-5 episodes of respiratory infections per year, like any other normal child. This has to be told to the parents and should not be equated to unsuccessful surgery.
4. Most children can begin or continue their routine immunizations about one month after their surgery.
5. Restriction of physical activities depends on the underlying uncorrected cardiac lesion and the treating cardiologist should be consulted in this regard. Children are good at limiting their own activity. A few weeks after discharge following cardiac surgery, the child can be allowed to play at his or her own pace if total repair had been done with no residual defect.
6. The child can attend the school after 2-3 days following percutaneous intervention (e.g. coil or device occlusion of patent ductus arteriosus, device closure of atrial septal defect) or after 1 month following the surgery with no residual defect or lesion (e.g. closure of atrial or ventricular septal defect). Those children with residual lesion can be allowed to attend the school after the first follow up with the cardiologist.
7. A hemoglobin level of 13-15 g% is desirable in all cyanotic patients and periodic iron supplementation is essential.
8. Pericardial collection can develop following discharge after cardiac surgery. This is particularly common after closure of ASD. The collection may occur as early as a few days following discharge.
9. The knowledge about the common cardiac medications and its timely modification is important. For example, the dose of most the drugs need to be increased when the child gains weight. Diuretics need to be withheld or reduced in the event of a child developing diarrhea.
10. Except patients operated for simple left to right shunts (ASD, VSD, PDA)all other children will need infective endocarditis (IE) prophylaxis for an indefinite period irrespective of the completeness of the repair.
11. Late unexpected death is a known complication in any complex congenital heart repair and the majority is due to arrhythmias. Checking the regularity of the rhythm is an important aspect of the follow up in these children.

Follow up has to be tailored to the individual patient.