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The Basis of Management of Congenital Heart Disease

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Additional information is available at the end of the chapter

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1. Introduction

1.1. Cardiac embryology — The development of the heart

1.1.1. Early heart development and the folding of the primitive tube

By the 3rd week of development the heart has developed from the cardiogenic region, a horseshoe-shaped structure, at the cranial end of the embryo, when it is about the size of a raisin. By day 21 the primitive heart tube has moved below the head region and by day 22 it fuses and moves into the future thoracic cavity and it is from this time that it begins to beat. The tube now starts to bend and twist and over the next 8 days, various chamber of the heart begin to develop and by the end of 2 months it bears a superficial resemblance to the fetal heart. [1]

The tube is anchored at one end by the arterial trunks and at the other end, by the various venous channels draining into it. Being fixed at both ends, the cardiac tube grows rapidly in length and begins to twist and bend. The embryonic ventricle is bent in to a loop to the right of the midline and the ventricle grows rapidly to cover the atrium and the great veins (figure 1). The sacculations projecting laterally will become the right atrium and the left atrium. The future left ventricle lies to the left of interventricular groove and the right ventricle or the bulboconus region communicates with the truncus arteriosus. A four chambered structure is formed from this convoluted tube by development of 3 septa which partitions the atria, ventricle and the truncus arteriosus. [2]

The septae develop simultaneously at about the same time between the 28 to 42nd day. The atria and ventricle are separated by a deep groove, the atrio-ventricular groove which appears like an invagination from inside. This forms the atrio-ventricular canal, which becomes divided

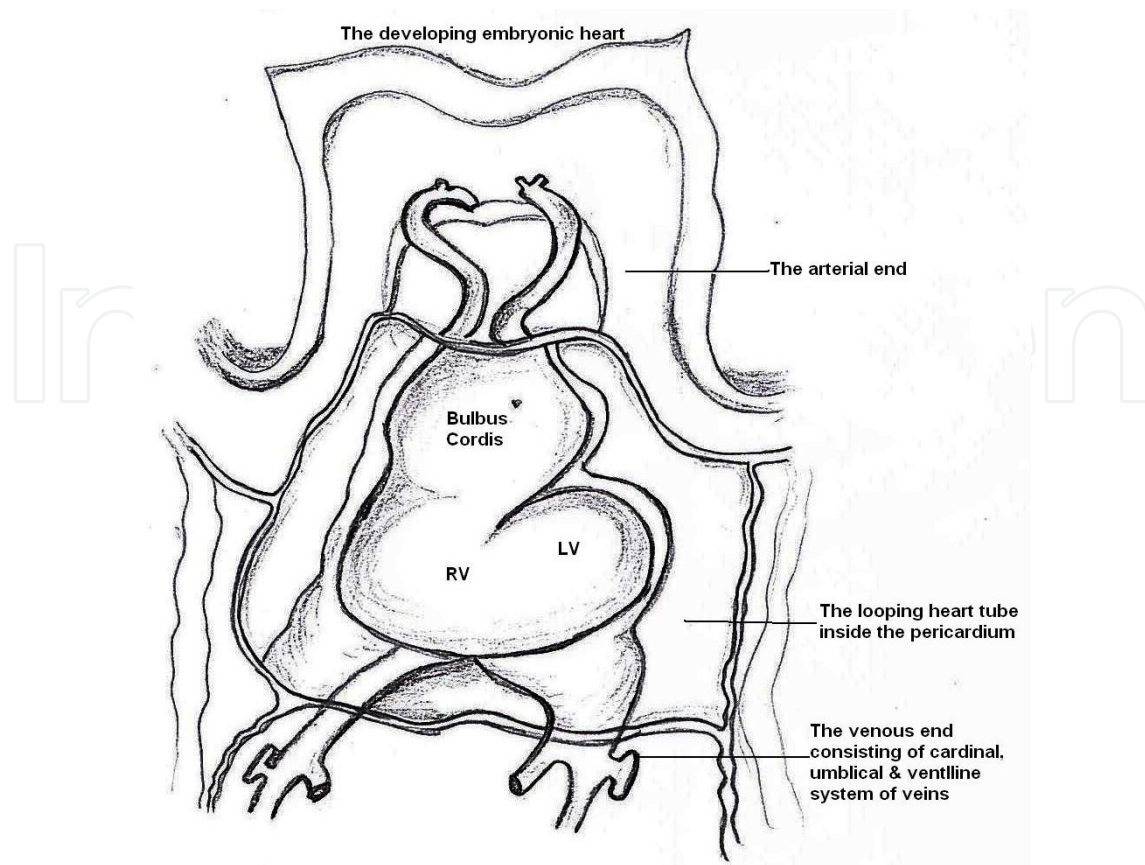


Figure 1. Development of the heart tube in the pericardial sac.

by the cushions which grow towards the junction. The endocardial cushions grow from opposite sides of the atrio-ventricular aperture and fuse to separate the atrium and ventricle.

From the interventricular ridge a proliferating muscular septum advances across the common ventricle towards the base of the heart. Simultaneously interatrial septum is formed by the septum primum growing rapidly towards endocardial cushions, leaving a foramen primum. Before the foramen primum becomes obliterated a new opening appears high on the interatrial septum- the foramen secundum- which allows shunting of blood from the right atrium to the left. The septum secundum develops from a ridge to the right of septum primum and grows like a curtain over the foramen secundum, the edge of the septum secundum forming the foramen ovale and the septum primum acting like a unidirectional valve, allowing blood to flow only from the right to the left.

The opening between the ventricular cavities -the interventricular foramen- persists, the closure of which depends on the development of a spiral septum which partitions the truncus and conus region into aorta and pulmonary artery.

The truncus arteriosus gives rise to the aortic arches, the 4th aortic arch forming the aorta and the 6th forming the origin of the pulmonary artery. A pair of ridges forms at the bifurcation which fuse and spiral down towards the interventricular foramen.

The interventricular foramen is obliterated by (a) masses of *endocardial tissue* at the interventricular septum, (b) masses of '*endocardial cushion*' tissue and (c) spiral septum. The partitioning of the heart is now complete. The aortopulmonary septum rotates 180 degree and fuses with the superior margin of the interventricular septum. This accounts for the manner in which the ventricular outflow tracts are aligned in a fully-developed heart- the aortic blood flowing posteriorly to anteriorly and the pulmonary blood flowing anterior to the aorta first and then posteriorly.

The events which occur during this period accounts for a majority of congenital heart disease. Atrial septal defect (which is most commonly a secundum defect) occurs due to defect of septum primum. Inadequate development of septum secundum (which forms by invagination of the developing superior vena cava and pulmonary veins) accounts for the sinus venosus type of defects.

The development of ventricular septum helps in understanding the predominance of perimembranous ventricular septal defects as three different regions have to fuse in a coordinated fashion to completely obliterate the interventricular communication. Uneven spiral partitioning of the outflow tracts can explain the occurrence of Tetralogy of Fallot (TOF) and double outlet right ventricle(DORV) and failure of the spiral pattern of aortopulmonary septum results in transposition of great arteries (TGA), when the aortopulmonary septum grows directly towards the interventricular septum. Failure of the septum to develop results in truncus arteriosus and the contribution of this septum to the interventricular septum explains the almost invariable association of truncus arteriosus defect with outlet VSD.

Failure of the endocardial cushions to develop properly results in the spectrum of endocardial cushion defects varying from ostium primum defects to partial and complete AV septal defects. [3]

1.1.2. Pulmonary venous development

Initially the blood coming from the lung buds drain into the splanchnic plexus which connects to the paired common cardinal and umbilicovitelline veins. The right common cardinal system forms the right SVC and azygous vein, the left common cardinal veins becomes the left superior vena cava and coronary sinus. The umbilicovitelline system becomes the inferior vena cava, ductus venosus and portal vein.

At 27 – 29 days the primitive pulmonary vein appears as an endothelial out pouching from superior left atrial wall and this joins the pulmonary venous plexus by 30 days. The common pulmonary vein enlarges and incorporates into the left atrium. The pulmonary venous part of the splanchnic plexus gradually loses its connection with cardinal and umbilicovitelline veins. Knowledge of the normal development of pulmonary venous pathway facilitates understanding of various types of anomalous pulmonary venous connections. [4]

1.1.3. Aortic arches

Aortic arches are series of six paired embryological structures connecting the ventral to the dorsal aorta. The ventral aorta at the level of 4- 6th arch fuses to form the truncus arteriosus

which forms the distal end of the developing heart tube. The dorsal aorta on the right usually disappears and the dorsal aorta on the left forms the descending aorta.

The first arch disappears and the 2nd persists as stapedial artery which is not of clinical significance. The third arch forms the internal carotid artery on both the sides and is called the carotid arch. The 4th arch on the right forms the right subclavian artery as far as the origin of the internal mammary branch and the left 4th arch forms the arch of aorta between the left carotid artery and the termination of ductus arteriosus. The 5th arch disappears on both sides. The proximal part of the right 6th arch forms the right pulmonary artery and the distal part disappears. The proximal part of the left 6th arch forms the left pulmonary artery and the distal part persists as the ductus arteriosus. (figure 2)

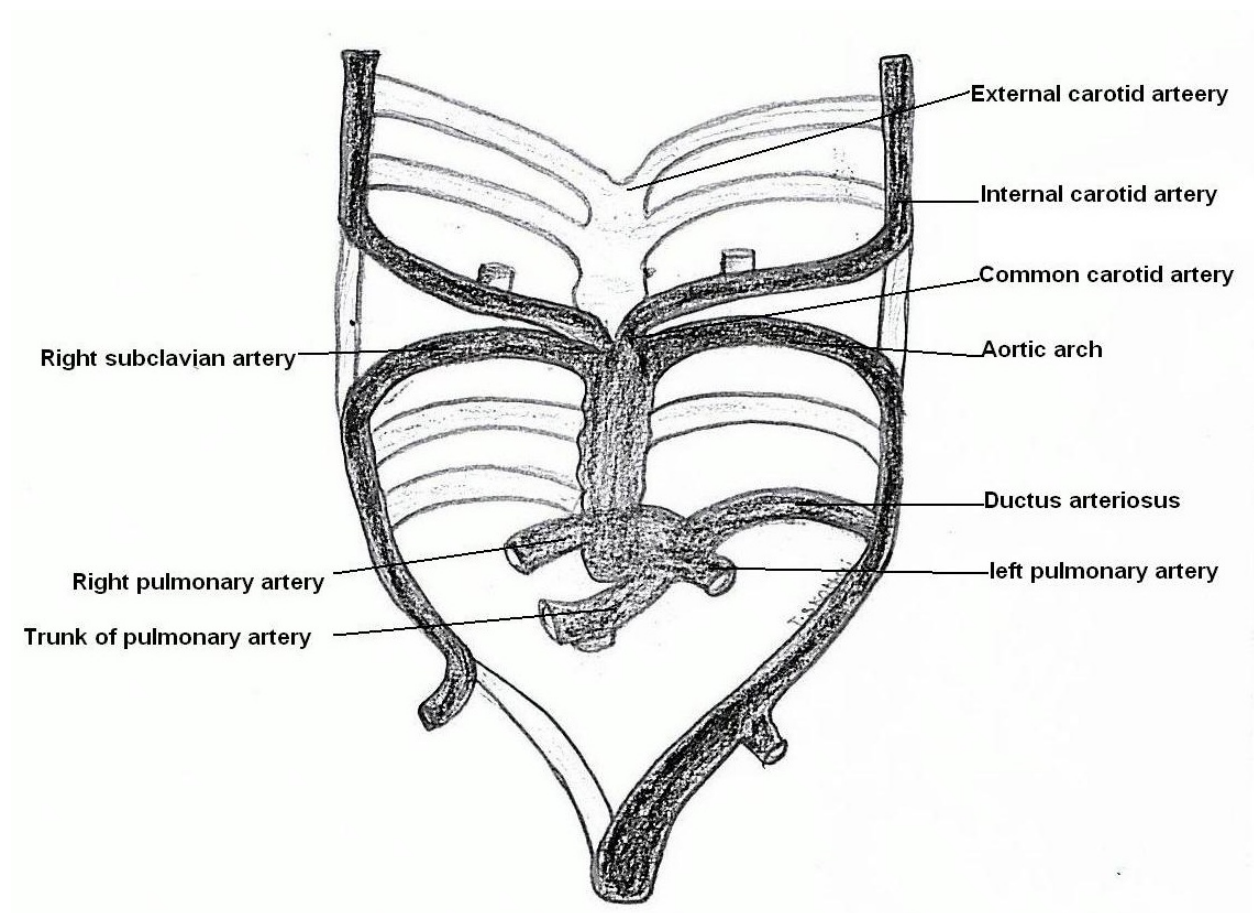


Figure 2. Development of aortic arches

Double aortic arch which is the commonest arch anomaly causing trachea and oesophageal compression occurs as a result of persistence of dorsal aorta. The recurrent laryngeal nerve loops around the 6th arch and hence goes around the ductus arteriosus on the left side and the subclavian artery on the right side. Persistence of the ductus arteriosus results in patent ductus arteriosus (PDA) and its excessive resorption can result in coarctation of aorta or stenosis of left pulmonary artery. [5]

The molecular mechanism behind the complex development is being now slowly unravelled. The consistent rightward looping of the heart suggests a highly conserved molecular control mechanism. The beating cilia on the node beats in a counter clockwise direction causing a leftward flow of fluid across the Hensen's node. The unidirectional movement is based on the inherent molecular chirality of the ciliary protein machinery. Pitx-2, a homeodomain containing protein plays a role in development of laterality. The Nkx2.5 and Tbx5 gene are expressed in atria and in conduction system. Irx4 is expressed only in ventricular chambers. The HAND 1 gene is expressed on the left ventricle and HAND 2 is expressed in the right ventricle.

The vertebral heart develops from precardiac mesoderm, anterior heart field and cardiac neural crest cells also play a critical part in myocardium and normal outflow tract development. The cardiac neural crest cell stabilizes the arch vessels and prevents their regression. A subpopulation of cardiac neural crest cells participates in the septation of outflow tract and contributes to the formation of semilunar valves. Retinoic acid is believed to have a role in signalling neural crest migration and cardiac development. Understanding the molecular and genetic mechanism may help in future fetal cardiac interventions. [6]

2. Surgical anatomy of the heart

The heart is enclosed in a pericardial sac. The pericardial cavity is the space between the inner lining of the fibrous pericardium and the surface of the heart. There are two recesses in the pericardial cavity lined by serous pericardium, the first is the transverse sinus behind the great arteries in front of the atria and is in free communication with the pericardial cavity on either side. The second is the oblique sinus, a blind ending cavity behind the left atrium.

The cardiac mass is 1/3rd to the right and 2/3rd to the left of midline. The ventricle is a three sided pyramid with diaphragmatic, anterior and left surfaces. The right sided margin is the acute margin and the left is the obtuse margin [7].

2.1. The morphological right atrium

The right atrium has 3 components-the appendage, venous sinus and the vestibule. (figure 3) The junction between the appendage and the venous sinus is marked by the prominent terminal groove. The groove internally corresponds to the crista terminalis, from which the pectinate muscle originates. The extensive array of pectinate muscles serves as one of the markers of the morphologic right atrium. Parallel and posterior to the groove is the second deeper groove between the right atrium and the right pulmonary veins. Dissection into this deep interatrial groove (Waterston's or Sondergaard's groove) permits incisions to be made into the left atrium (the *classic posterior approach* to the left atrium).

The sinus node lies in the subepicardial position at the cranial part of the terminal groove, and is a spindle-shaped structure which lies lateral to the superior cavoatrial junction. The artery to the sinus node arises from right coronary artery (55%) or circumflex coronary artery (45%).

The septum between the right and left atria is formed by the floor of the oval fossa and its adjacent anteroinferior muscular rim. The superior rim or the septum secundum is formed by deep interatrial fold extending between the systemic and the pulmonary veins. Larger part of the anterior atrial wall is related to the aortic root (the torus aorticus). It is important to realize that the true atrial septum is rather small and it is easy to go outside the heart when attempting to gain access to the left atrium through the septal approach. Because of the infolding of the interatrial groove, access to the left atrium can be gained by approaching through the right atrium and incising superiorly within the fossa (*The septal superior approach*). [8]

The triangle of Koch which contains the atrioventricular (AV) node is an area of major surgical significance. The triangle is demarcated by the (a) the tendon of Todaro, (b) the attachment of the septal leaflet and (c) the orifice of the coronary sinus. The tendon of Todaro is formed by the fusion of the valve guarding the IVC (the Eustachian valve) and the coronary sinus (the Thebesian valve). This inserts into the central fibrous body and runs in the tissue separating the oval fossa from the mouth of the coronary sinus. The trabeculated diverticulum found posterior to the coronary sinus is called the post-Eustachian sinus of Keith.

The node lies in the atrial muscle above the hinge point of the septal leaflet, and the bundle which arises from the node penetrates the interventricular septum at the apex of the triangle of Koch.

The central fibrous body is actually the area where the membranous septum and leaflets of the atrioventricular and the aortic valves join in fibrous continuity.

2.2. The morphological left atrium

Like the right atrium, the left atrium has a (a) venous sinus (b) appendage and (c) vestibule. The differences however are

- a. The venous component of the left atrium is considerably larger than the appendage
- b. the junction between them is NOT marked by a terminal groove or crest.
- c. The pectinate muscles are confined within the appendage and do not extend around the vestibule
- d. The appendage is long, tubular and finger like with a narrow end unlike the broad triangular appendage of the right atrium

Among these the confinement of the pectinate muscles within the appendage is the most reliable differentiating factor between the morphological right and left atrium.

2.3. The morphologic right ventricle

It has three components- the inlet, trabecular and outlet parts.

The inlet portion is limited by the tricuspid valve and its tension apparatus

The trabecular component extends to the apex, where it is thin and it is vulnerable to perforation by cardiac catheters and pacemaker electrodes.

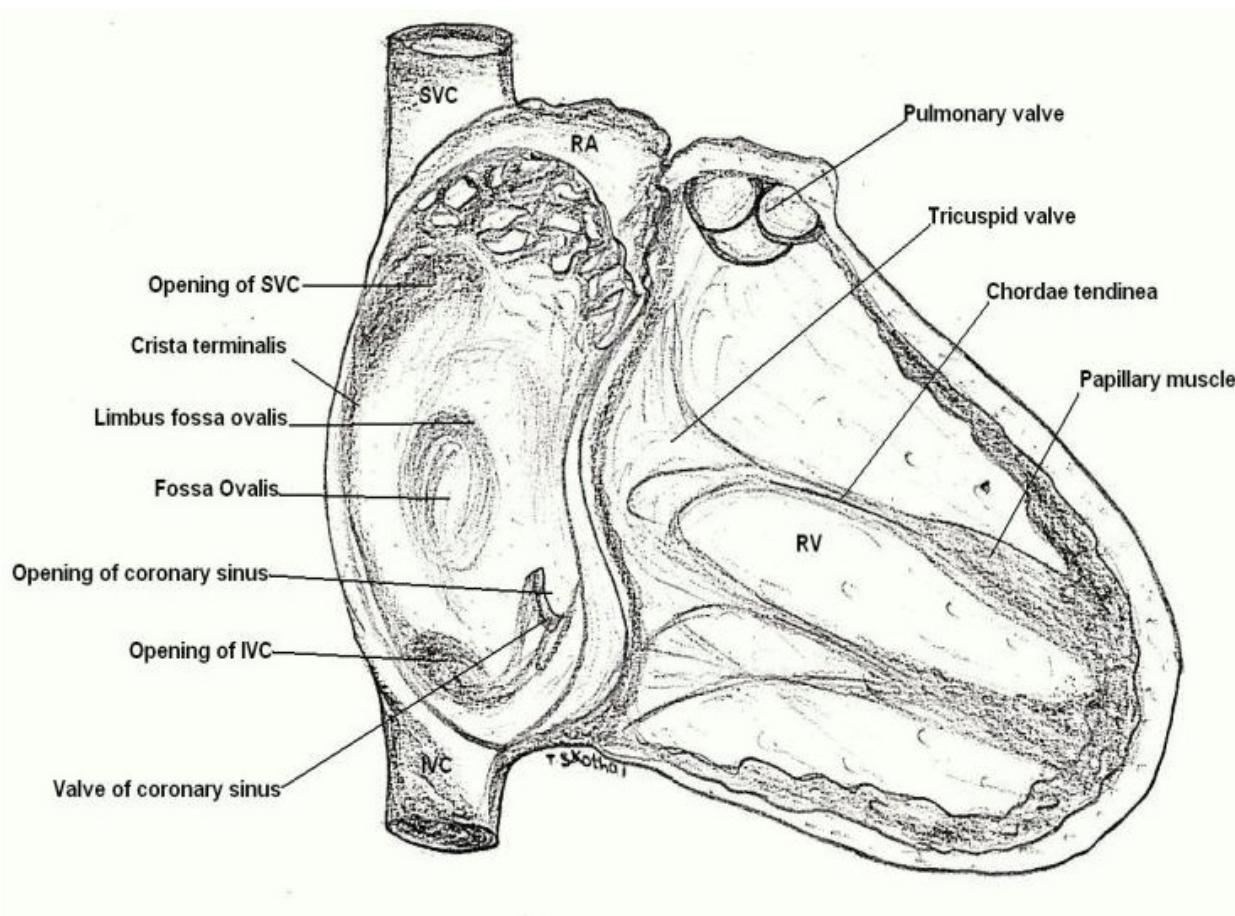


Figure 3. Right atrium and ventricle

The outlet component of the right ventricle is a complete muscular structure - the infundibulum- which supports the pulmonary valve.

The muscular shelf separating the tricuspid and the pulmonary valve is the supraventricular crest. Much of the crest is no more than the infolded inner heart curve and incisions and sutures deep in this area can jeopardize the right coronary artery. The distal part of the crest is continuous with the sub-pulmonary infundibulum, the presence of which permits the pulmonary valve to be removed and used as autograft during the Ross procedure.

The crest is cradled between two limbs of the prominent right ventricular trabecula called the septomarginal trabeculation, which has a superior limb, an inferior limb and a body.

The superior limb runs up to the attachment of the pulmonary valve.

The inferior limb gives rise to the medial papillary muscle (of Lancisi) and a line drawn from this muscle to the apex of the triangle of Koch marks the position of the atrioventricular conduction axis.

The body runs to the apex, and gives rise to the moderator band and the anterior papillary muscle.

The coarseness of the apical trabeculation is the most constant feature of the right ventricle. The other differences are the direct septal attachment of the tension apparatus of the atrioventricular valve, which is usually tricuspid in nature.

2.4. The morphologic left ventricle

The inlet component is limited by the mitral valve and its tension apparatus. The mitral valve has two leaflets, the aortic or the anterior leaflet which is in fibrous continuity with the aortic valve, which is short and square and the other leaflet which is connected to the wall of the left atrioventricular junction is called the mural or the posterior leaflet.

The leaflets do not have direct septal attachment unlike the right ventricle but instead attach through anterolateral and posteromedial papillary muscles.

The apical myocardium is thin like the RV. The septum is not completely muscular unlike RV, and it has a small membranous part which forms the subaortic outflow tract.

The muscular septal surface is characteristically smooth and the left bundle lies below the membranous septum corresponding to the zone of apposition between the right coronary and non-coronary leaflets.

The semilunar valves of the aortic and pulmonary valves are similar, the distal portion forms the sinotubular junction and the proximal part takes origin from the ventricular structure. The overall arrangement is like the crown, rather than forming an annulus. [9]

The aorta and pulmonary artery form the vascular pedicle. The aorta gives rise to the brachiocephalic or innominate artery, the left common carotid artery and the left subclavian artery. The pulmonary artery arises anteriorly and courses posteriorly, and is a short vessel giving rise to the right and left pulmonary arteries. The left pulmonary artery lies superior to the left bronchi in front of the descending aorta. The right pulmonary artery is anterior to the left main bronchus and has a long mediastinal course beneath the aortic arch and behind the superior vena cava to reach the hilum of the right lung. Sometimes an early branching large upper lobar branch can be mistaken for the right pulmonary artery.

The coronary arteries arise from the aortic sinuses. According to Leiden convention the position is described in terms of an observer from the non-coronary sinus, the right hand of the person is called the sinus 1 and gives rise to the right coronary artery and left hand facing sinus is called the sinus 2 and gives rise to left coronary artery. The coronary artery arises beneath the sinotubular junction, and when it is displaced more than 1 cm from the ST junction it is considered abnormal which occurs in 3.5% of hearts.

The left coronary artery has a single orifice, while in 50% there are two orifices in the right sinus, one gives rise to the main RCA and the smaller orifice gives rise to the infundibular or sinus nodal artery. It is important while giving ostial cardioplegia for stopping the heart to perform cardiac surgeries to instill into the smaller orifices to protect the sinus node.

The epicardial course of the coronary arteries follows the atrioventricular and interventricular grooves. The RCA gives to the acute marginal branches, the sinus nodal artery (55%), and

posterior descending artery (90%) at the crux which supplies the diaphragmatic surface. The LCA gives rise to the anterior interventricular (left anterior descending) and the circumflex branches. The anterior interventricular artery gives rise to the diagonal branches to the obtuse surface of the heart and the septal perforating branches. The circumflex artery gives rise to the obtuse marginal branches of the heart. [10]

The Coronary veins accompany the artery and drain into the coronary sinus.

The Great cardiac vein runs along the left anterior descending artery and encircles the mitral orifice, and at its left margin receives the oblique vein of the LA and forms the coronary sinus. The coronary sinus lies between the left atrium and left ventricle before joining the right atrium.

The Middle cardiac vein runs along the posterior descending artery and the small cardiac veins accompany the right coronary artery. The Thebesian valve guards the orifice of the coronary sinus.

3. Conduction system and its surgical significance

3.1. The development

The heart develops from the mesoderm from fusion of vascular channels which forms the heart tube. The heart starts beating from the time the embryo is 22 days old. Even from the beginning there is a polarity and the peristaltic type of motion of the tube starts from the venous end and ends in the arterial end. This sequential contraction ensures that even in the absence of valves there is very little regurgitation of the blood, and an ECG similar to the adult ECG is recognized at the end of 1st month. It is believed that this primitive heart tube persists as the remnant of conduction tissue and the myocardium grows around this to form the atrium and the ventricle. The conduction system is one of the most primitive structures in the heart which forms even before the heart tube starts looping.

There is a circle of conduction tissue at the atrioventricular junction, which as the atrioventricular valves form and the great arteries are assigned to the respective ventricles gradually becomes restricted. The only area of electrical continuity between the atrium and the ventricles is the AV node and the penetrating bundle where the muscular septum comes in contact with the AV junction. This establishes connection with the Purkinje network in the ventricular myocardium, to ensure sequential contraction of atrium and ventricles. (figure 4)

This sequence of events occurs in the usual d-looping of the heart. If there is l-looping of the heart as in congenitally corrected transposition, where the right atrium joins the left ventricle, which then gives rise to the pulmonary artery and the left atrium joins the right ventricle which gives rise to aorta, the bundle can be extremely elongated bringing it under the pulmonary valve anteriorly. The conduction system is vulnerable to injury during surgery and otherwise. Both congenital and spontaneous heart blocks can occur at the rate of 2% per annum in this condition. [11]

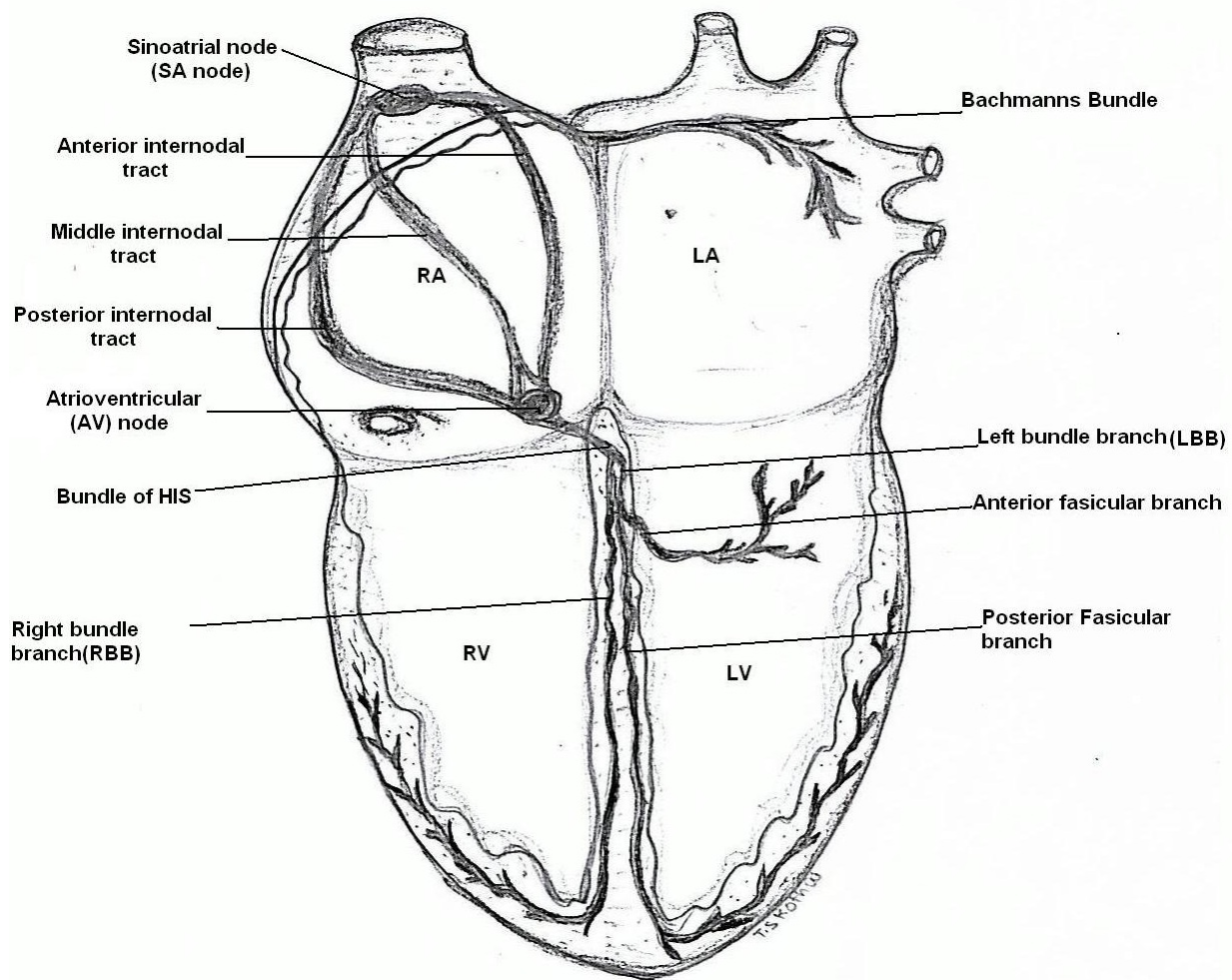


Figure 4. Conduction system

3.2. Surgical perspective

3.2.1. Sinoatrial (SA) node

SA node which is located at the lateral cavoatrial junction at the upper end of crista terminalis is the pacemaker which initiates contraction. This can be identified as a small oval shaped structure which is slightly more yellowish with SA nodal artery forming small ramifications over it.

This area is vulnerable during –

1. Glenn surgery – where the SVC is divided and anastomosed to the pulmonary artery end to side – the cavopulmonary anastomosis. Care has to be taken while clamping and dividing the SVC.
2. Sinus venosus atrial septal defect with partial anomalous pulmonary venous connection – some of the pulmonary veins can drain high into the SVC and during correction

sometimes it may be necessary to cut across the cavoatrial junction longitudinally which makes the SA node or the artery supplying this at risk.

3. Sennings procedure –the SA node is at risk during suturing the pulmonary venous baffle.
4. Superior septal approach to the mitral valve – which may put the artery supplying the SA node at risk. Injury is manifested by nodal rhythm during which the P wave is absent in the ECG. The distal portion of the conduction system gradually takes over or the SA node itself recovers, making the need for a permanent pacemaker rare in this setting.

3.2.2. Atrioventricular (AV) node

The AV node lies in the apex of the triangle of Koch, the penetrating bundle pierces the apex of the triangle and reaches the crest of the trabecular septum immediately beneath the membranous septum which is below the junction formed by the *right and noncoronary cusp*. It is in this area that the bundle is most vulnerable during surgical closure of ventricular septal defect. This corresponds to the commissure between the anterior and septal leaflets of the tricuspid valve which is usually supported by the medial papillary muscle. Sometimes there is a cleft in the septal leaflet which points to this area.

The penetrating bundle crosses from the atrial septum to the aortic outflow and then joins the muscular septum. In normal hearts with normal alignment the apex of the triangle is close to the crest of the interventricular septum. The length of the bundle is small in this setting and this length of the non-branching bundle can be excessive in the presence of VSD in the inlet portion of the ventricular septum.

It is only when the ventricular septum reaches the crux of the heart can a regular AV node join the AV conduction axis. The node and bundle is formed at the place where the ventricular septum joins the AV junction.

A few rules may help avoid heart block while closing VSD's

- a. In perimembranous defects, which is also the type of defect usually seen in tetralogy of Fallot, the VSD patch should be sutured about 5 mm away from the posteroinferior margin, and in perimembranous defects with outlet extension the muscle protects the bundle and makes it more left sided and this makes the posteroinferior angle safe in VSD with outlet extension. In the perimembranous defects with inlet extension where the bundle is superficial and close to the posteroinferior angle of the defect, and hence is exposed to maximum risk of damage. The risk of damage to the node is minimal in cases of muscular and subpulmonic VSD's.
- b. The base of the septal leaflet tissue is always safe to place sutures.
- c. The fibrous tissue surrounding the VSD can be used to anchor the patch and this has to be differentiated from aneurysm of membranous septum which may sometimes harbour the bundle.

- d. In AV canal defects the AV node and the coronary sinus are displaced inferiorly, and the AV node is placed in between the coronary sinus and the crest of interventricular septum at the so called 'NODAL triangle' rather than at the usual apex of the triangle of Koch. [12]

Transient damage to the conduction system can occur as a result of myocardial protection during cardiopulmonary bypass, which should usually recover in less than 7 days. A permanent pacemaker is usually needed if sinus rhythm does not return in 9-10 days

4. Fetal and neonatal circulation

The circulatory system evolved as the simple process of diffusion of nutrients from amniotic sac is no longer able to meet the metabolic needs of the growing embryo.

The placenta does the function of oxygenation in the fetus as the lungs are bypassed by presence of ductus. The arrangement of circulation in the fetus is such that the more oxygenated blood goes to the head and the less oxygenated blood goes through the ductus into descending aorta and into the umbilical arteries.

The umbilical vein brings the oxygenated blood which bypasses the liver through the ductus venosus and this blood coming into the IVC is preferentially streamed by the Eustachian valve into the left atrium through the foramen in the atrial septum and this oxygenated blood enters the left ventricle and from there to the aorta and arch vessels.

The more deoxygenated blood from the SVC goes into the right ventricle which is pumped by the RV into the PA and as the fetal lungs are collapsed with very high resistance in the pulmonary circulation, the blood bypasses the lungs and goes into the descending aorta through the ductus.

A few lesions provide insights, which can evolve into more complicated congenital heart pathologies. Absence of foramen is one such lesion, which has been linked to the development of hypoplastic left heart syndrome, the development of which can be prevented by dilating the foramen ovale.

Atresia of the pulmonary and aortic valve can also be intervened by ballooning to prevent the development of its sequel which can be hypoplastic left heart and RV dependent coronary circulation in cases of pulmonary atresia with intact ventricular septum.

Absence of ductus arteriosus is a condition which is associated with absent pulmonary valve. The absence of ductus could be the primary condition which leads to RV output regurgitating back into ventricle due to high fetal pulmonary vascular resistance, with the VSD partially decompressing the right ventricle. The increased right ventricular output causes the main and the branch pulmonary arteries to dilate. This could extend into the lungs, compressing the airways, and thereby causing severe respiratory compromise in neonatal period as seen in extreme cases of TOF with absent pulmonary valve.

5. The transition from fetal to neonatal circulation

The first breath causes the lungs to expand and oxygen content in the blood to increase and this provides the impetus for the ductus to constrict and close. The right ventricular output goes to the lungs and reaches the left atrium, the increased pressure in the left atrium causing the flap valve to shut, closing the foramen ovale. The umbilical vein and ductus venosus regress to leave the vestigial ligamentum teres and the ligamentum venosum.

Maintaining the patency of ductus is critical to many potential fatal neonatal conditions. These can be any of the conditions in which the output of the heart through either the aorta or the pulmonary artery is critically reduced and the patent ductus maintains the flow from one great artery to the other. These are hypoplastic left heart syndrome, critical aortic stenosis and pulmonary atresia. The presence of ductus is also useful in mixing lesions like TGA though the degree of mixing is much better in the presence of atrial level communication. Patency of ductus can be maintained by Prostaglandin E1 infusion, the availability of which has enabled the stabilization of many sick neonates before subjecting them to surgery [13].

6. The pathophysiology of L->R shunt lesions

ASD, VSD and PDA are the main lesions which shunt from left to right.

In Atrial septal defect, the size of the shunt is determined by the size of the defect and the degree of pulmonary vascular resistance. Small shunts cause no enlargement of cardiac chambers, while large shunt cause significant RA and RV dilatation, mid-diastolic flow murmur though the tricuspid valve and ejection systolic murmur at the left upper sternal border due to increased flow across the pulmonary valve. The left chambers do not increase in size as the increased return to LA is decompressed into the RA through the defect.

The pathophysiology of the shunt in VSD and PDA are similar. The magnitude of shunt is determined by the size of the defect and the degree of pulmonary vascular resistance (PVR); PVR is more important in the large defects where it determines the degree of shunt. Since PVR is elevated at birth and falls by 6-8 wks, children with large VSD typically become symptomatic with signs of congestive heart failure (CHF) at this time.

A small defect causes no enlargement of cardiac chambers, the ECG and X-ray are normal, there is ejection systolic murmur and P2 is normal. For moderate sized defects, there is LA and LV enlargement unlike ASD and as the RV is contracting at the time of left to right shunt, it undergoes no volume load. However the increased blood pumped by the RV causes mid-diastolic murmur across the mitral valve.

Large defects and PDA cause CHF in infancy, especially if the PVR is low. This reflects in biventricular hypertrophy with increased saturations, pan systolic murmur and loud P2. As the PVR increases, the heart size becomes normal on Chest X-ray though the pulmonary segment remains prominent, the murmur is reduced, and there is pure RVH in ECG

In PDA in addition there is enlargement of aorta and transverse aortic arch, which is usually not very evident in on X-ray as the aortic arch does not form a part of cardiac silhouette. As the PVR increases, there can be differential cyanosis with lower limb saturations being lower than the upper limb reflecting right to left shunt at the level of PDA.

Endocardial cushion defects present with features of both ASD and VSD. The QRS axis is abnormal between -20 to -150 degree, which is due to the disposition of the His bundle and its branches intrinsic to the pathology and not due to hemodynamic consequence.

Obligatory shunts are those where the degree of shunting does not depend on the PVR, examples of which is Gerbode defect where there is LV to RA shunt or ruptured sinus of Valsalva [14]

7. Stenosis and regurgitation of valves

If there is stenosis of atrioventricular valves, it causes systemic or pulmonary venous congestion, depending on the valves involved. Regurgitant lesions cause volume loading of both upstream and downstream chambers. Unlike stenotic lesions which cause pressure overload, regurgitation lesions cause volume overload.

Aortic regurgitation causes wide pulse pressure and the regurgitating jet causes diastolic flutter of mitral valve causing the Austin Flint murmur, and the jet also causes the mitral valve to close early and reduces the intensity of S1. Aortic regurgitation in children usually occurs as a part of ventricular septal defect when the leaflets either attempt to close the defect or get sucked in due to Bernoulli effect, or prolapse due to lack of support to the leaflets due to absence of continuity of media.

Pulmonary regurgitation is common after TOF repair and the significance of which depends on the diastolic distensibility of the right ventricle: patients with diastolic dysfunction of the RV tolerate PR better with less dilatation, and narrow QRS. TR causes prominent liver and neck pulsations.

7.1. Cyanotic heart defects pathophysiology – TGA and Truncus arteriosus

TGA is one of the common cyanotic conditions presenting in infancy where the aorta arises from the RV and PA arises from the LV. The systemic blood reaches the RV and from there to aorta and again back to RV, (the blood flow is parallel rather than in series). Such an arrangement is incompatible with life in the absence of atrial, ventricular or ductal level communication. In the presence of inadequate communication, they can present with very poor saturations (30-50%) with acidosis and hypoglycaemia in the first week of life. Of all the levels of communication the atrial septal defect provides the best area of mixing without streaming and atrial septostomy (Rashkind procedure) done for this purpose is the first palliative intervention for a congenital heart disease, which started off the practice of pediatric interventional cardiology.

Any newborn with deep cyanosis and cardiomegaly (egg on side appearance) and increased pulmonary vascular marking and no murmur can be diagnosed to have TGA. Patients with VSD present a little late with features of early CHF and these patients are prone to develop very early pulmonary hypertension, because they get relatively desaturated blood under high pressure. The unique feature of pulmonary circulation of 'hypoxic vasoconstriction' accelerates the onset of pulmonary vascular disease. TGA, VSD and PS can occur and the presentation of which depends on the degree of reduction of pulmonary blood flow.

Truncus arteriosus has complete mixing of systemic and pulmonary blood and the level of saturation is proportional to the degree of pulmonary blood flow.

7.2. Pathophysiology of Tetralogy of Fallot

This is commonest cause of cyanosis in children in developing countries. Though classically supposed to have the features of a) VSD b) Aortic over-ride c) RV hypertrophy and d) Right ventricular outflow tract (RVOT) obstruction, the two important features are

- a. A large VSD at least as big as the aortic annulus to equalise pressures in both ventricles.
- b. Right ventricular outflow tract obstruction – which can be at any level infundibular, valvar, supra-valvar or at branch PA level.

If a child presents clinically with signs of small VSD with RVH, it strongly suggests diagnosis of TOF, the probability of which increases in the presence of right aortic arch.

The intensity and the duration of heart murmur are inversely proportional to the severity of pulmonary stenosis. In pulmonary atresia or in cyanotic spells when there is critical reduction in pulmonary flow there may be no murmur at all due to absence of flow across the RVOT

There are no signs of CHF in TOF because no chamber is under volume overload and only RV is under pressure overload which is not suprasystemic and is well tolerated.

The degree of cyanosis depends on the balance between the systemic and pulmonary resistances, and decrease in SVR due to activities like crying and defecation can increase the degree of R->L shunt.

The role of RVOT spasm in the development of cyanotic spell is controversial

Hyperpnoea plays an important role in the perpetuation of cyanotic spell as it increases the venous return and more desaturated blood enters the systemic circulation due to override.

Termination of a spell can be achieved by

- a. By increasing SVR – by knee chest position, phenylephrine, ketamine
- b. Reducing hyperpnoea by sedation – morphine, sodium bicarbonate which reduces respiratory stimulation, ketamine
- c. Decreasing venous return – squatting
- d. Stabilisation of vascular reactivity – propranolol prevents sudden decrease in SVR and its role in preventing RVOT spasm is controversial.

7.3. Pathophysiology of Tricuspid atresia and TAPVC

This is a single ventricle physiology where there is enlargement of RA, LA and LV and hypoplastic RV. 70% have normally related great arteries and 30% have transposed great arteries. In either of these situations the pulmonary blood flow can be increased or decreased. QRS axis is deviated leftward with LVH and the axis resembles endocardial cushion defect.

TAPVC – Total anomalous pulmonary venous connection can present as supracardiac, cardiac and infracardiac. The timing of clinical presentation depends on the presence of obstruction.

Non-obstructed TAPVC presents like large ASD. Obstructed variant can present with extremely sick child with pulmonary venous congestion causing ground glass appearance on chest X-ray, severe cyanosis, respiratory distress and severe pulmonary arterial hypertension. Murmur is usually soft or absent.

Any child with features of pulmonary oedema and ground glass appearance on chest X-ray with normal size cardiac shadow and no murmurs should be considered to have obstructed TAPVC. [15]

8. Chest X-ray

X-ray is integral part of the evaluation of a child with cardiac disease.

The x-ray helps in the evaluation of cardiomegaly, chamber size, and blood flow by looking at the pulmonary artery and venous markings. Lungs, spine, thorax and visceral situs are also evaluated using x-ray. For example the presence of aortic knuckle and gastric fundus on the same side is suggestive of corrected transposition.

The structures forming the margin of the heart on the right side are– SVC, aortic knuckle and RA and on the left side they are the pulmonary artery, left atrial appendage and LV.

Different conditions can have diagnostic X-ray features. TOF has a '*boot shaped*' heart, TGA has a '*egg on side*' heart and supracardiac TAPVC is associated with '*snow man sign*' the left vertical vein, the innominate vein and the right superior vena cava form the head of the snowman., Truncus, the dilated pulmonary artery particularly the right pulmonary artery produces the '*comma or the water fall sign*'. In Ebsteins anomaly there is cardiomegaly with a narrow pedicle, with '*Pencil line sharp*' cardiac borders. These classic appearances are not usually seen, though they are supportive evidences in broader clinical context.

8.1. The assessment of pulmonary arterial and venous pressure using X-ray

Pulmonary plethora – the presence of right descending pulmonary artery larger than the size of trachea is a sensitive sign of increased pulmonary blood flow. Other signs are prominent upper and lower zone vessels and vessels seen in the outer third of the lungs. Infants and children present with generalized mottling, due to increased pulmonary flow.

Pulmonary arterial hypertension: Is said to occur when the mean pulmonary artery pressure is > 20 mmHg,

In mild PAH – (20-29 mmHg) there is prominent pulmonary artery.

Moderate PAH- (30-49), the central vessels dilate further,

Severe PAH (50 mmHg) - there is central dilation with reduction in the calibre of peripheral vessels (peripheral pruning). The size of the pulmonary artery correlates with PAH, and it has been found that plethora correlates better with the degree of left to right shunt than cardiomegaly.

Pulmonary venous hypertension – normal 8-12 mm Hg, and gradual increase causes:

- a. Redistribution of blood flow (10-20mm Hg) – Cephalization due to dilation of upper zone vessels and blurring of lower zone vessels
- b. Interstitial oedema – (15-25 mm Hg) Kerley lines and peribronchial cuffing
- c. Alveolar oedema- (25 – 35 mm Hg) causes the 'Bat's wing appearance' when the interstitial fluid accumulates at rate faster than it can be removed by lymphatics.

The presence of prominent aortic knuckle points to the presence of extra cardiac left to right shunt like PDA, Sinus of Valsalva rupture, Coronary arteriovenous fistula, or AP window. The absence points to intra- cardiac L-> R shunt like atrial septal defect and ventricular septal defect.

In addition to the above findings careful consideration should be given to lung parenchyma to look for any parenchymal patches and also the status of spine and bony thorax for complete preoperative assessment.

9. Echocardiography

Using ultrasound to visualize organs was first introduced in the 1970's and over the 1980's it transformed the field of imaging becoming the primary diagnostic modality for evaluation of congenital heart disease. During the 90's and 2000's steady progress has been made in the areas of 3D imaging, myocardial function assessment and trans-esophageal echocardiography(TEE).TEE is now routinely used intraoperatively for planning and performing cardiac procedures, TEE probes can now be placed in children as small as 3.5 kg.

Echo is non-invasive, has excellent spatial and temporal resolution, ability to see the anatomy and physiology in real time along with portability. Echo is now everywhere right from prenatal imaging, preoperative, intraoperative, postoperative and follow-up imaging.

A burst of ultrasonic energy is produced by the piezoelectric crystals placed on the transducer probe which passes through the tissue and the returning ultrasound is processed by amplification, filtering and is analysed to display in a moving real-time format.

The different modes of imaging are

- a. M- Mode – uses a narrow ultrasound beam to provide a ‘ice pick’ image of the structure. It has good axial resolution. Used to measure the degree of movement of leaflets. Chamber thickness is measured using 2-D directed M-mode imaging.
- b. 2-D mode –uses *phased array transducers* which are multiple piezoelectric crystals that both transmits and receives ultrasound simultaneously; used to provide an image of the ‘section’ of the heart.
- c. 3-D mode *uses matrix array transducers* and sophisticated parallel array processing to provide real time image. With progressive miniaturization, real-time 3D Transesophageal images provide excellent images intraoperatively for planning valve repairs.
- d. Doppler imaging – can be used to estimate the velocity and direction of blood flow to estimate the pressure gradient cross sectional flow area and prediction of intracardiac pressures.
- e. Contrast Echocardiography – is based on the fact that any intravascular injection produces a contrast which can be detected by echocardiography. Used for intracardiac and great artery level shunts in patients with poor windows, for detecting pulmonary venous malformation and for detecting baffle leak following atrial switch procedures.
- f. Fetal echocardiography – using trans abdominal screening majority of the cardiovascular malformations can be detected by 17- 20 weeks of life; by transvaginal window, heart and great vessels can be visualized at the end of 1st trimester. [16]

10. Magnetic Resonance Imaging

It is a imaging modality which uses magnetic fields and radiofrequency energy to stimulate hydrogen nuclei which emits radiofrequency waves that are used to construct images. Over the 1990’s the field has evolved from a procedure which takes a long time to produce a series of static images to one in which real time 3-D visualization is possible. MRI uses magnetic field of strength 0.5 Tesla to 3 Tesla (1 Tesla is 10,000 Gauss; earth magnetic field is 0.5 G). MRI uses fields which are 5000 to 60000 more powerful than the earth’s magnetic field.

Synchronisation or respiratory motion compensation is required as the heart is a moving object and ‘gating’ or ‘synchronization’ is required to return to the same point in the cardiac cycle in order to freeze the cardiac motion. Pulse oximetry, ECG signal or MRI navigator echoes can be used for gating. Advances in *gradient coil and parallel acquisition* methods can obviate the need for synchronization.

Two main modalities of MRI are:

- a. Spin Echo – uses a radiofrequency pulse that tilts the hydrogen protons by 90 degrees followed by a second 180 degree pulse, which are used to generate images. Produces images in which the flowing ‘blood is black’. This provides static anatomic information, with excellent blood myocardium contrast. Used for assessing cardiac tumours, pericardial disease and thoracic masses. Takes a relatively longer time

- b. Gradient Echo - sequences uses radiofrequency pulses that are less than 90 degrees, that are faster than spin Echo images and used to produce images in which the flowing 'blood appears white'. 'Steady State free precession' MR sequences allow real time MR fluoroscopy, with shorter imaging times. Used to assess ventricular function and flow of blood in the cardiac pathways and for identifying stenotic or regurgitant jets.

Myocardial tagging – uses 'spatial modulation of magnetization' so that protons in selected volume are incapable of producing a signal. This produces stripes across the image, tagging the myocardium. As the heart moves the tags are followed and this allows calculation of myocardial strain

Velocity encoded Cine MRI- can be used to measure blood flow velocity and quantify blood flow rate.- can measure the regurgitation volume and can even calculate the shear stress exerted by the blood on the vessel wall.

Contrast enhanced MRI – Uses gadolinium chelate which produces bright blood signal used for clear delineation of spatial relationship and for imaging of baffles and outflow tracts. Hyper enhancement of myocardial regions observed 10-15 minutes after administration of gadolinium contrast is indicative of scar tissue and irreversible myocardial injury.

MRI in patients younger than 5-6 years would require sedation, Surgical clips, sternotomy wires, coils stents and occluding devices are MRI safe once the surrounding fibrous tissue grows over these implants and makes them immobile. Cardiac pacemakers, presence of intracranial, intra ocular or intracochlear implant are considered contraindication to MRI.

Cardiovascular MRI is fast becoming a tool which can provide us with anatomic and functional information not provided by echo or cardiac catheterization. [17]

11. Cardiac catheterisation and interventions

Catheterisation used to be the main diagnostic modality available when it was first introduced in 1946. The era of angiographic anatomic delineation is fading. Echocardiography is now preferred for evaluation of valvar and congenital cardiac defects with 3D echo promising real time surgical images of the valves for repair. MRI and CT angiography is fast replacing angiogram for delineation of complex relationship, volume estimation of chamber, extra cardiac vessels, aortic arches and venous anomalies. The field of MR imaging, has the potential to completely replace diagnostic angiography. The routine use of catheterization before single ventricle surgeries is being questioned, as the same information can be made available through non-invasive means

Angiogram still has a role in

- a. Visualising branch PA anatomy beyond hilum- in case of tetralogy of Fallot with aortopulmonary collaterals
- b. Coronary anatomy.

- c. Measurement of flow, pressure, reactivity and resistance of pulmonary vasculature to various drugs in patients with elevated pulmonary artery pressure.
- d. Estimation of cardiac output, Q_p/Q_s calculation for assessment of operability.

The cardiac output can be calculated by the thermodilution technique or the Fick's principle.

Fick's principle is based on the fact that

'The uptake or release of substance by an organ is equivalent to the blood flow to the organ multiplied by the arteriovenous difference of the substance'

O_2 is used as the indicator and all the systemic organs or lungs are considered as one organ to estimate the systemic or pulmonary output respectively.

We use the O_2 consumption in ml/min/m² and this divided by the arteriovenous difference would give the systemic or pulmonary flows. The consumption is made available through estimates based on age, sex and heart rate.

Systemic output would be the difference between the arterial oxygen content and mixed venous oxygen content (ideally measured in the middle of right atrium to average for the superior vena caval, inferior vena caval or coronary sinus venous blood whose oxygen content may be different)

Pulmonary output would be the difference between the pulmonary venous and pulmonary artery oxygen content.

Q_p/Q_s can be easily calculated even if we do not have the values of oxygen consumption. We need the systemic arterial, mixed venous and pulmonary arterial and pulmonary venous oxygen content (which is assumed to be 100% in the absence of pulmonary pathology).

Pressure measurements are based on fluid filled catheters, or large bore catheters with multiple side holes which would yield accurate description of intracardiac waveforms. The resistance is calculated using the Ohm's law, which is the difference in the pressure across the organ divided by the amount of blood flowing through it. PVR (Pulmonary vascular resistance) would be transpulmonary gradient (difference between the mean PA and LA pressures) divided by the pulmonary flow. Measured in mmHg/ L/min/m² or Wood units [18]

12. Interventions

With the availability of modified catheters and catheter delivery devices, the role of intervention in pediatric cardiology is increasingly becoming important

Catheter interventions can primarily do three things

1. Open things that are closed: Atrial septostomy – done in infants who need mixing of blood for maintaining saturation and cardiac output. Most common indication is transposition of great arteries with intact ventricular septum, other rarer indications are TAPVC with

restriction at the interatrial septum, tricuspid atresia, pulmonary atresia, mitral atresia. Septostomy with a blade would be required if the infant is > 6-8 weeks as the septum becomes thicker.

2. Widen things that are narrow – involving the blood vessels and heart valves, done using balloon catheters and stents to prevent recoil in case of vessels. The stent gets incorporated into the vessel wall. There can be neointimal proliferation causing restenosis, usually in the first 2 years. The procedure uses special plastic polymer balloons which will not inflate beyond predetermined size even under high pressure. This needs a guide wire to be placed across the narrow area with balloon placed across so as to place the waist (middle of the sausage) in the narrow region which is dilated with dilute contrast.

Used commonly for

- a. Pulmonary valvar stenosis- for gradients > 40 mmHg, has almost replaced surgery, except when the valve is very dysplastic or associated infundibular obstruction
- b. Peripheral and branch pulmonary artery stenosis in the setting of postoperative tetralogy of Fallot, pulmonary atresia,- are surgical challenges and balloon dilatation with stenting has emerged as preferred alternatives with 60-80% success rate.
- c. Aortic valve stenosis – used when gradient > 50-60 mm Hg in the absence of AR can be life-saving in small infants, can also be used for discrete subvalvar aortic stenosis but not for tunnel like subaortic stenosis.
- d. Coarctation of aorta – preferred for re-coarctation or for coarctation in grown up children where it can be combined with stenting. Not for coarctation in infancy which has high rate of recurrence and surgery is better option.
- e. Mitral valves – works well for rheumatic heart disease, not very successful with congenital MS. Heart block, tearing of leaflet and occurrence of MR are possible complications.
- f. Prosthetic valves and conduits
- g. Systemic vein stenosis in the setting of post- operative Mustard or Senning's procedure, does not work in the presence of pulmonary vein stenosis.
- h. Close things that are open – this uses devices mounted on catheters that are passed through long and large sheaths after crossing the area to be closed.

ASD, VSD and PDA's – ASD's upto 32 mm diameter can be closed, the defect needs a rim of 4 mm for the device to be centred and placed. Amplatzer devices are FDA approved and have the longest track record.

Closure of VSD requires careful assessment to ensure that the device would not interfere with tricuspid or aortic valvar mechanisms. Arrhythmias, stroke, perforation, device embolization, incomplete closures are the risks involved.

Large PDA's > 5mm are closed with Amplatzer I device which has a single aortic rim and is mushroom shaped.

Coils – are metal wires coated with thrombogenic Dacron strands, suitable for vessels < 6-7 mm, with an area of narrowing. The thrombus formation around the coil plugs the vessel. Embolization, incomplete closure and hemolysis are possible risks.

Other defects which have been plugged are ruptured sinus of Valsalva especially the ones arising from the non-coronary sinus, aorta LV tunnels, veno-venous collaterals plugging following cavopulmonary connection. [19]

13. Pediatric anesthesia and critical care

The success of any pediatric cardiac surgical programme depends on a team effort and anesthetists and intensive care personnel are critical members of the team. The importance of effective communication between the various team members cannot be overemphasized.

Care of pediatric patients and care of the neonates in particular needs appreciation of the differences in **physiology** of the immature organ systems.

The neonate responds rapidly to any stressful situations which reflect in the sudden changes of various metabolic and hemodynamic parameters. The metabolic rate of neonates is 2-3 fold have increased compared to adults. The caloric requirement is between 100-150 Kcal/kg/day and neonatal gut is at risk of necrotizing enterocolitis with the use of hyperosmolar feeds.

The myocardium has only 30% contractile tissue as against 60% in mature myocardium. The myocardium has a reduced ability to respond to afterload stress and the compliance is reduced. Acute pressure afterload is poorly tolerated, and can lead to rapid ventricular dysfunction; chronic pressure load is longer tolerated than chronic volume load. Symptoms of CHF are rare unless the obstruction is severe and prolonged.

The stroke volume is relatively fixed and cardiac output is more heart rate dependent. They are more dependent on the trans-sarcolemma movement of calcium to initiate and sustain contraction as the sarcoplasmic reticulum and T-tubules are relatively underdeveloped.

Cardiorespiratory interactions and ventricular interdependence are particularly marked in infants. Positive pressure ventilation reduces the preload of both ventricles, increases the afterload of right ventricle and reduces the afterload of left ventricle. Implying that in situations where right ventricle is dysfunctional or in situations of single ventricle, early extubation with reduced afterload would be useful and in situations of left ventricular dysfunction, positive pressure ventilation would reduce the afterload of the heart and act as positive inotrope. This principle should be tempered against the background of ventricular interdependence where dysfunction of one ventricle can rapidly affect the other due to septal interactions.

Neonates rely on diaphragm as the main muscle of respiration. Only 25% of the diaphragm in neonates is made of type I fibre capable of slow and sustained activity against 55% by 8 months of age. Raised intra-abdominal pressure due to any cause like gastric distension, hepatic congestion and ascites can compromise its function. In neonates a larger portion of energy expenditure is used for ventilation and therefore they fatigue easily and have failure to thrive

in the presence of increased work of breathing. They have increased closing capacity with airway closure occurring during normal tidal ventilation putting them at risk of developing hypoxemia and atelectasis. In addition dilated pulmonary arteries and left atrium can compress bronchi causing lobar collapse. [20]

Pulmonary hypertension both pre and postoperatively plays an important role in the planning of surgery, anesthesia and postoperative care.

Preoperatively it can be due to large left to right shunt lesions, or due to pulmonary venous obstruction, rarely due to pulmonary vascular obstructive disease.

Intraoperatively- light anesthesia, hypoxemia, hypoventilation, lung hyperinflation or hypoinflation, hypothermia, respiratory and metabolic acidosis, protamine, blood products, prolonged bypass with inflammatory response and capillary leak, compression and atelectasis of lung, pulmonary edema from inadequate venting of left atrium can all contribute to increased pulmonary vascular resistance (PVR).

Intravenous drugs can be used to reduce the PVR, but they lack selectivity and can cause systemic hypotension. Nitroprusside, glyceroltrinitrate, milrinone, Prostaglandin E1 and I2, tolazoline and isoproterenol have been used. Inhaled NO is most selective pulmonary vasodilator currently available; it is rapidly taken up and inactivated by haemoglobin as it diffuses from the alveoli. Oral drugs in the form of PDE type V inhibitor, sildenafil and endothelin I blocking drugs bosentan have shown encouraging results. [21,22]

Preoperative evaluation should keep in mind the physiology of defect, and the changes that the preoperative treatment could have caused (diuretics causing hypokalaemia) and the presence and severity of cyanosis and pulmonary hypertension. Haematocrit greater than 65% can exacerbate tissue hypoxia and can cause stasis and potential thrombosis. Avoiding dehydration is very important to avoid tissue hypoxia and to maintain renal function postoperatively.

Non-invasive monitoring using electrocardiography, pulse oximetry, capnography, non-invasive blood pressure is placed before induction, invasive arterial and central venous line should be planned according to procedure. Neurologic monitoring and cerebral protection is of concern during congenital heart surgery. Nasopharyngeal temperature, continuous EEG, transcranial Doppler, frontal lobe infrared spectroscopy and cerebral oximetry can be used to evaluate cerebral blood flow velocity and perfusion. Intraoperative echocardiography has achieved a significant role in repair of CHD. It helps in re-evaluation of anatomy before intervention, adequacy of surgical repair and de-airing after weaning from cardiopulmonary bypass and has become an integral part of monitoring in many units including ours.

Maintenance of diastolic pressure and coronary perfusion is important particularly in the setting of duct dependent lesions and in situations of altered coronary perfusion. There is a choice of induction techniques, inhalational, IV or IM.

Fentanyl (15-25 ug/kg), ketamine (1-3 mg/kg), Pancuronium (0.2mg/kg) or Suxamethonium (2mg/kg) in combination with glycopyrrolate (10ug/kg) allows prompt induction and airway control without significant increase in PVR. Midazolam (0.1-0.2mg/kg) is also a useful adjunct

during narcotic induction but can cause hypotension in patients with high sympathetic drive. Isoflurane and midazolam can be used during bypass for maintenance and blunt awareness.

Reducing stress response using high dose opioid anesthesia and extending this to immediate postoperative period was considered important to reduce morbidity and mortality. With changes in surgical practice and particularly the timing of surgery, a strategy of using high dose opioid may be a less critical determinant of outcome. The main aim however is to maintain hemodynamic stability so that the team can focus on surgery without the distraction of side effects of anesthetic drugs.

14. Discontinuation of cardiopulmonary bypass and postoperative management

A co-ordinated approach must be used during weaning so that a smooth transition can be ensured - there should be close communication between surgeon, anesthesiologist and perfusionist. The need for vasopressors and inotropes is decided by close observation of the heart during re-warming period. Once adequate temperature is reached, ventilation is restarted after a good inflation of the lungs to prevent atelectasis and for deairing. Acid base status is normalized, adequate heart rate is ensured and the drainage from venous cannula is slowly reduced and CPB is stopped.

Myocardial edema and secondary fall in cardiac output by 20-30% is common in neonate in the first 6 – 12 hrs following surgery. Sternal closure is frequently delayed in neonates due to this reason. Avoiding hypothermia and hypoglycaemia, maintenance of optimal filling pressures, and preventing abdominal distension due to ascites by using catheter which can be used for peritoneal dialysis are important in the early postoperative period. An LA line and PA line may be inserted by the surgeons in the theatre in situations when LV dysfunction is anticipated or when pulmonary artery pressures are labile. This would help to fine tune management and help in decision making with regards to inotropes and ventilation management.

Postoperative management requires a precise knowledge about the anatomy, pathophysiology and details of the surgical and CPB technique. For most patients the recovery is uncomplicated and whenever the clinical progress does not follow the expected course possible residual or additional defects should be investigated.

14.1. Analgesia and sedation

Inadequate analgesia while the patient is on ventilator may be manifested by tachycardia, hypertension, pupillary dilation, diaphoresis etc, Changes in the respiratory pattern like tachypnea, grunting and splinting of the chest wall may also be seen. Fever, hypoxemia, hypercapnia, seizures, vasoactive infusions, early low cardiac output may also present in similar fashion and should be ruled out.

Opioid analgesics are the mainstay of pain management as they blunt hemodynamic response to procedures such as endotracheal suctioning. Morphine in intermittent or continuous infusion (50-100 ug/kg) is an excellent analgesic with sedative property. Disadvantage is that it can cause histamine release with systemic vasodilation and elevation of PVR. Fentanyl (5-10ug/kg/hr) is an alternative drug with less sedative action, and does not cause histamine release. There is wide variation in the metabolism of fentanyl, tolerance and dependence develops rapidly and chest wall rigidity can develop as a rare idiosyncratic reaction. It blocks the stress response and maintains systemic and pulmonary hemodynamic stability.

Dexmedetomidine (commonly called the dexmed) is alpha-2 agonist which is increasingly being used due to its sedative, anxiolytic, and its non-respiratory depressant property. This can cause hypotension and bradycardia and should be used with caution in children with CHD.

Recognition and early intervention for the management of low cardiac output is one of the pivotal roles of the intensivists. The following are some of the clues and the entire clinical picture should be considered rather than an isolated finding.

Physical examination: Core hyperthermia, tachycardia, cool peripheries with impalpable peripheral pulses, hypotension with narrow pulse pressure, ascites, hepatomegaly, oliguria, obtundation of sensorium.

Monitoring – Dampened arterial upstroke, narrow pulse pressure, elevated venous pressures (systemic or pulmonary – loss of sinus rhythm, residual outflow obstruction, tamponade, AV valve regurgitation should be ruled out)

Laboratory – Metabolic acidosis, low mixed venous oxygen saturation (or increased arterio-venous oxygen difference > 25 – 30%), increased arterial lactate, potassium, liver transaminases and Increased BUN and creatinine.

Strategies for management of Low CO should focus on optimizing the balance between oxygen supply and demand.

Demand – Maintaining adequate analgesia, sedation and paralysis when necessary, strict avoidance of hyperthermia and occasionally using mild hypothermia to reduce metabolic rate.

For optimizing delivery – Oxygen content can be optimized by managing Hemoglobin and f_iO_2 , and the factors which determine output

Contractility – Dopamine (5-10ug/kg/min), dobutamine (5-10 ug/kg/min), epinephrine (upto 0.1ug/kg/min is considered acceptable. Requirement greater than 0.3 – 0.5ug/kg would make one assess the possibility of mechanical circulatory support. Calcium infusion may also be necessary especially in patients with diGeorge syndrome and 22q11 deletion.

Afterload – Milrinone (0.2- 0.75 ug/kg/min) has inotropic and peripheral and pulmonary vasodilating property and is useful for patients with ventricular dysfunction and increased afterload. Nitroprusside and GTN can be used in the setting of normal ventricular function. Norepinephrine (upto 0.2ug/kg/min) and vasopressin (10-120 milliunits/kg/hr is a potent vasopressor) can be used in situations with low SVR ('warm' shock) [23]

Heart rate and sinus rhythm –temporary atrial and ventricular pacing wires are kept after any major cardiac procedure and can be used to optimize the heart rate, though a little trial and error would be required to find the best settings for the individual patient

Stress dose steroid – hydrocortisone (50mg/m²/day) has been demonstrated to increase systemic blood pressure and lower inotropic scores and should be used when the pressures are recalcitrant to inotropes. No consistent correlation has been found between the serum cortisol level and low cardiac output state. Due to risks of infection and poor wound healing its use should be restricted to 3- 5 days.

T3 at dose of 0.05ug/kg/hr has been shown to be beneficial in neonates due to sick euthyroid state when there is reduced conversion of thyroxine to active T3.

Fluid management – the first 24 hrs the maintenance fluids should be 50% of full maintenance and volume replacement should be titrated to filling pressures and hemodynamic response. Furosemide (0.2-0.3 mg/kg/hr is used after iv bolus of 1 mg/kg can provide consistent and sustained diuresis. Fenoldopam is a new selective dopamine DA1 receptor agonist with renal and mesenteric vasodilating properties (0.1-0.5 ug/kg/min).

Peritoneal dialysis, hemodialysis and continuous veno-venous hemofiltration provide renal replacement therapy for patients with persistent oliguria and renal failure. Besides helping in solute and water clearance, they help in nutritional support by allowing additional volume, and may also remove the inflammatory mediators. [24]

Weaning from ventilation and extubation should be possible once patients are hemodynamically stable and normothermic with good cardiac output. Patients after closed heart surgery like PDA ligation, ASD, small to moderate VSD, RV to PA conduit change, bidirectional Glenn shunt and Fontan procedure with fenestration are suitable for early extubation. Neonates undergoing surgery like TGA, Truncus, TAPVC, TOF would require elective ventilation for 48 hrs for the physiology to normalize.

Extubation of an infant after a major cardiac procedure is both an art and science. Upto to 25% of 1st extubation can end in reintubations, and if more than 3 attempts fail serious consideration should be given to the following conditions. Residual volume and pressure load, ventricular dysfunction. Phrenic nerve injury, bronchomalacia, retained secretions, vocal cord injury, Diuretic therapy with contraction alkalosis, Inadequate nutrition and an evolving sepsis are all factors to be considered for failed extubation. Early tracheostomy can sometimes help if there is no correctable lesions, and if predominantly retained secretions and nutrition are the major causes of failed extubation. [25,26]

14.2. Cardiopulmonary bypass and extracorporeal life support

Cardiopulmonary bypass machine essentially takes over the function of pumping and oxygenating the blood allowing surgery to be performed on arrested still heart.

Gibbon developed the first cardiopulmonary bypass machine used for closing an atrial septal defect in 1953, Throughout the 1960's the typical survival rate for open heart surgery remained around 50%. During the 1970's the mortality of CPB was reduced by using deep hypothermic

circulatory arrest pioneered by Barrat- Boyes and Castaneda. Progressive improvement in circuit design and perfusion techniques has now brought the overall mortality to less than 5%. Even now the morbidity associated with the use of CPB is held to be the major limitation for completely successful outcomes.

Due to immature organ function, fetal contractile protein isoforms, immature calcium cycling, pulmonary dysfunction and exaggerated stress and inflammatory response the morbidity associated with CPB is more profound in infants and neonates.

The Cardiopulmonary bypass circuit consists of

1. Oxygenators - microporous membrane(0.05 – 0.25um) pores which allows more efficient oxygenation but lasts shorter are used for cardiac surgery, the nonporous folded sheet silicone membrane oxygenator is usually selected for longer term circulatory support applications.
2. Pumps – Roller pumps are used and the flow rate is governed by the revolutions per minute, occlusion produced by the rollers and the internal diameter of the tubing.
3. Tubing – usually ¼ inch tubing is used on arterial and venous limbs, the pump is situated close to the field to reduce the tubing length which contributes to the priming volume.
4. Venous reservoirs – Open rigid reservoirs are usually used to which the blood flows by gravity, the level of blood in the reservoir is an important safety mechanism – source of volume for arterial inflow and to judge adequacy of venous return. Malposition of venous cannulas and lost blood in the surgical field can reduce the venous volume which may need urgent attention. Collapsible venous reservoir with reduced air blood contact is increasingly being used. Vacuum can improve drainage through small venous cannulas and tubing which can theoretically reduce organ edema and improve organ function; venoarterial air embolism is a potential complication.
5. Arterial cannula – aorta is the usual site of cannulation, the location needs to be tailored according to surgery. Alternate sites are carotid artery in small infants and femoral artery in bigger children (> 10-15kg).
6. Venous cannula – SVC and IVC cannulas are used to maximize venous return and to minimize interference with the operative field, and optimally sited to prevent obstruction and kinking.
7. Filters – 0.2 um filters are used in gas inflow and for crystalloid and cardioplegia solutions. 40um filters are used in cardiotomy suction return lines to remove macro and micro aggregates and debris returning from the surgical field. Arterial filter (40um) may reduce microemboli, though some consider its use optional.
8. Prime – The fluid used to fill the oxygenator, minimum level required in the reservoir and the tubing is the priming volume. The prime consists of physiologic crystalloid solutions, packed red cells, colloids like albumin and FFP, and mannitol (used for its membrane stabilization, antioxidant and osmotic diuretic properties). The hematocrit during CPB should be maintained around 25–30 %. At low temperature and low flows, low hematocrit

may improve microvascular flow and oxygen delivery. For patients with myocardial dysfunction and complex prolonged surgeries a target of around 40% at the end of surgery may improve oxygen delivery.

14.3. Initiation, monitoring and termination of CPB

Heparin at a dose of about 4mg/kg is given to bring activated clotting time (ACT) of more than 400 seconds. ACT should be maintained between 400- 600 seconds to prevent activation of blood coagulation and clot formation. Inadequate concentration of heparin is believed to contribute to excess coagulation and fibrinolytic system activation.

Once the arterial and venous cannulae are in place, after confirming adequate anticoagulation and absence of air (especially at the arterial cannula and tubing) is confirmed, CPB is slowly initiated by beginning arterial inflow and unclamping the venous line. Any systemic to pulmonary shunts should be closed prior to, or immediately after CPB initiation. These can contribute to systemic runoff, contributing to organ malperfusion, increased left heart return, heart distension, and inadvertent rewarming. It is absolutely essential to prevent myocardial distension at all times after initiation of CPB.

The important CPB circuit variables that are monitored are:

1. Arterial line pressures (typically in the range of 200-250mm Hg to drive blood through the infant arterial cannula and tubing to maintain mean pressures around 40-60 mm Hg).
2. Pump flow rate - which is a calculated value in roller pumps based on rotation, internal tubing diameter and occlusion pressure.
3. Oxygenator gases – the flow rate ('sweep speed') and oxygen concentration controlled with blender and flow meter, usually started with 1: 1 ratio with the flow rates in membrane oxygenators.
4. Temperature- thermistors measure the temperature of the water bath, arterial and venous blood. The gradient between the patient and the perfusate should not exceed 10 degree C, especially during rewarming to prevent formation of gaseous bubbles.

Hypothermia delays loss of ionic hemostasis, slows consumption of ATP, decreases free radical generation, inflammatory cytokine production, white cell activation and leucocyte adhesion molecule synthesis, suppresses the release of excitatory amino acid neurotransmitters. It continues to be the mainstay for cerebral and other organ protection during CPB.

The important patient variables monitored are

1. Patient mean arterial and venous pressures – femoral arterial line preferred especially in small infants and in deep hypothermia.
2. Nasopharyngeal (reflecting brain temperature), rectal or bladder (core) and esophageal (aortic) temperatures are measured using appropriate thermistors, slow cooling over a period of 15-20 minutes- is necessary whenever deep hypothermic circulatory arrest (DHCA) is planned. Hct of 25% is favoured at this temperature.

3. Arterial and venous blood gases are measured every 15-30 minutes. Oxygen saturation of venous blood (SvO₂) and blood lactates are an important index of tissue perfusion and SvO₂ can also be measured continuously using calibrated inline monitor. Values < 60-70% should raise concerns of inadequate tissue oxygen delivery. At low temperature, due to increased affinity of Hb to oxygen, higher levels (90%) may have to be targeted
4. Blood glucose should be monitored frequently particularly in neonates and infants who are prone to hypoglycemia due to low glycogen reserves.
5. pH management – There are two methods –
6. Alpha stat - maintains electrical neutrality at lower temperatures, pH measured is 7.7 at 20° C. Advantages are better preservation of metabolic functions and buffering capacity, useful during **mild and moderate** hypothermia.
7. pH stat - Adds CO₂ to the circuit to correct pH for the fall of temperature, useful during **deep** hypothermia to increase cerebral blood flow and decrease metabolic rate, and also for management of aortopulmonary collaterals to increase pulmonary vascular resistance and to increase systemic flow.

15. Bleeding and organ injury

The coagulation factors reach the adult level at 6 – 12 months of age. The effects of CPB on blood activation and coagulation is far greater in neonates and infants because of hemodilution, hypothermia, greater shear stress, and more blood- air contact activation.

Platelets are the initial therapy for bleeding after adequate heparin reversal. 1 unit of platelets per 10 Kg raises the platelet count by approximately 50,000/mm³. Cryoprecipitate which is a good source of fibrinogen is the next blood component usually used. Antifibrinolytic agents like aprotinin, e-aminocaproic acid, tranexamic acid have become popular to reduce bleeding after complex surgeries and in infants.

Lung, kidney and brain are at risk of injury during CPB. The stress and inflammatory response is 5-10 times greater in neonates and infants. High dose steroids have been used to mask the response and have been shown to be more beneficial in neonates and infants. Ultrafiltration, both conventional and modified are being used for hemoconcentration, removing inflammatory mediators, and decreasing total body water, with beneficial effects being shown for hematocrit, oxygenation, pulmonary vascular resistance and decreasing duration of mechanical ventilation.

16. Extracorporeal life support

Modalities include extracorporeal membrane oxygenation (ECMO), intra-aortic balloon pump (IABP) counter pulsation and ventricular assist devices. Wide application in pediatric age

group is limited by the need for miniaturization and ECMO is still the most common form of mechanical circulatory support for pediatric patients.

ECMO is now accepted modality of treatment in neonates with a variety of parenchymal and vascular lung disease (meconium aspiration, diaphragmatic hernia, persistent hypertension of newborn). The outcome irrespective of the indication depends on *early diagnosis, prompt institution and reversible nature of dysfunction*. The use of ECMO for respiratory indications has progressively decreased (due to increased availability of High frequency oscillatory ventilation, nitric oxide, and surfactant therapy) and there is a steady increase in its use after congenital cardiac surgery or as a bridge to transplantation to support failing circulation.

The cardiac indications for ECMO are in preoperative resuscitation (critical AS, pulmonary hypertension, obstructed TAPVC, transposition of great arteries), inability to wean from CPB, cardiomyopathy, myocarditis, after in-hospital cardiac arrest and CPR, and bridge to transplantation.

Venovenous ECMO is used in patients who need ventilatory support alone. Arteriovenous cannulation is used for cardiac ECMO. In the postoperative period single right atrial and ascending aortic cannula is preferred, internal jugular vein and carotid artery can be used in other settings. ACT is maintained around 180-200 seconds, flow rates between 100-150 ml/kg/min for children on full ECMO, hematocrit level between 35-45% and platelet count > 100,000/mm³.

The daily management of ECMO needs assessment of cardiorespiratory function, end organ perfusion and evolving complications such as bleeding or sepsis. Assessing adequacy of flow and systemic perfusion is of paramount importance.

When instituted in post cardiac surgery setting, myocardial recovery should be anticipated in 2-3 days, failing which listing for transplantation or withdrawal of support must be considered. Patients with cardiomyopathies or with severe bronchiolitis due to viral infection may require longer period of support (1-2 weeks).

Weaning depends on the underlying indication. Inotropes are recommenced or increased, intravascular status is optimized and ventilator settings are adjusted, flows gradually decreased over a period of time and the circuit is clamped. ABG, serum lactated levels and mixed venous oxygen saturation levels are closely monitored and decannulation is done once patient has maintained stable circulation and acceptable gas exchange for upto 4hrs.

IABP (Intra-aortic Balloon Pump) has not been very successful in paediatric population because of a number of reasons, the usual right heart nature of pathology, rapid heart rate in children making timing difficult, distensible nature of aorta and collaterals making coronary flow augmentation and afterload reduction less. The smallest size available is 2.5ml and generally volume of 0.5ml/kg is recommended. Technical improvements including smaller sized consoles, catheters and M-mode echocardiography for timing may enhance its applicability in the future.

Ventricular assist devices require direct cannulation of heart. Reported indications are decompensated cardiomyopathy, ALCAPA (ischemic myocardium as a result of anomalous

origin of coronary artery), and retraining of poorly prepared left ventricle after arterial switch procedure. They are simple in design and require less technical assistance once established. VAD are more suited as bridge to transplantation. Berlin Heart Excor VAD is the first extracorporeal pneumatically driven pulsatile VAD designed specifically for paediatric use and is available in 6 different sizes depending on stroke volume. Significant advances in the circulatory support technology for children are expected in the near future with various devices being in preclinical stages of development. [27]

17. Conduits in cardiac surgery

Allografts – tissues obtained from human body which are cryopreserved are used in repair of variety of congenital cardiac defects. Due to availability and preservation problems it fell into disrepute in 1980's only to re-emerge in 1990's mainly due to late complications of porcine valved Dacron conduits which became popular in 1980's and availability of better cryopreservation technology.

Allografts are currently available standards against which any conduit is compared. Factors which can reduce the longevity of allografts are – use of excessively large conduits ($> +2$ z score), aortic allografts which tends to calcify earlier probably because of higher elastin content (controversial) and immunogenicity due to blood and tissue incompatibility (controversial). No particular method is proven to enhance the longevity of allograft. Decellularisation process using anionic detergents and nucleases have been used to provide acellular matrix for the native tissue to repopulate and grow, but the long term structural impact of the decellularisation process is yet to be seen. Treating the decellularised tissue in a bioreactor using the recipient cells so as to repopulate the matrix is now in experimental stages.

Contegra which is made from Bovine jugular vein is one of the popular valved conduits in pediatric cardiac surgery; This conduit shows predictable function with survival approaching homografts over mid-term. The advantages are easy availability (12- 22mm sizes) and predictable quality. It is important to keep the conduit short and straight to prevent 'telescoping' and distal stenosis.

Alternative conduits have been used made of Polytetrafluoroethylene (PTFE) wall and leaflets, which has minimal tissue reactivity. Long term results are still awaited.

Conduits fashioned out of autologous pericardium with PTFE (0.1mm) leaflets are also increasingly being used, the disadvantages being non-availability in reoperations and limitation with respect to the length and diameter.

While a variety of choices are available for older children and adults, an ideal conduit for neonates and small children is still elusive. The availability of 'conduits which grow' using tissue engineering technology can potentially provide curative surgery for a number of conditions which require multiple reoperations for conduit replacements. [28]

18. Surgical approaches to the correction of congenital cardiac defects:

A simplified approach to such a variety of congenital cardiac defects is to broadly classify them as

1. left-to-right shunts
2. cyanotic conditions
3. single-ventricle physiology
4. valvular and coronary heart disease
5. tracheal and vascular rings

18.1. Left to right shunts

Left to right shunts are the most gratifying to treat as they are potentially life-saving and promote the growth of the child. The common lesions are atrial septal defect (ASD), ventricular septal defect (VSD) and patent ductus arteriosus (PDA).

The common variations of ASD [29] (figure 5) are ostium secundum, ostium primum, sinus venosus, coronary sinus and various combinations of these. Secundum ASD's are usually closed by the interventional cardiologists percutaneously provided parameters are satisfied including adequacy of rims and the safety of neighbouring structures. When these criteria are not met, surgery is via median sternotomy or right posterolateral or anterolateral thoracotomy (cosmetic), placing on cardiopulmonary bypass (CPB) and closure either directly or using the patient's own (autologous) pericardial patch. Ostium primum ASD is usually associated with abnormal mitral and sometimes tricuspid valves. Repair involves repair of the valves with closure of the ASD with a patch. Failure to close the cleft in the mitral valve increases reoperation rate for mitral regurgitation [30]. These patients require lifelong follow-up for mitral regurgitation and left ventricular outflow tract obstruction [31] Complete heart block is a potential complication due to the proximity of the bundle of His.

Sinus venosus ASD is usually superior vena cava (SVC) ASD with partial anomalous pulmonary venous connection (usually right upper lobe pulmonary veins draining anomalously to the right atrium instead of left atrium). Surgical options range from simple ASD closure (single-patch), two-patch technique and Warden's procedure which is more complex [32]. Complications range from SVC obstruction, pulmonary venous obstruction and sino-atrial nodal dysfunction. Coronary sinus ASD is rare and involves patch repair. Common atrium is complete absence of interatrial septum and surgery is partitioning of the atria. Autologous pericardial patch is the material of choice.

Ventricular septal defect (VSD) is deficiency of the interventricular septum. There are various classifications based on anatomy and embryology. The commonest classification is perimembranous VSD, muscular VSD, doubly committed subarterial VSD, inlet septal and multiple VSD's. [33] The majority close by one year, particularly perimembranous and muscular. The doubly committed and inlet septal VSD's do not close spontaneously. Indications for surgery

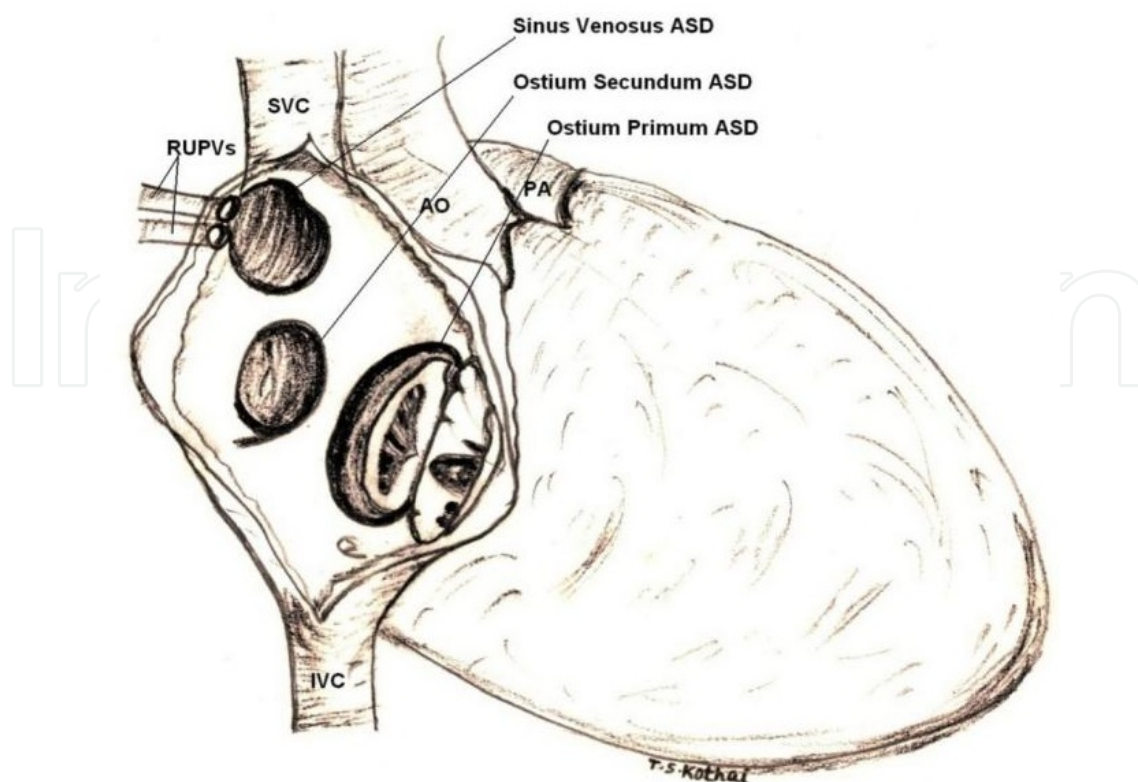


Figure 5. Atrial Septal Defects

are large VSD with congestive cardiac failure refractory to medical management, left atrial and left ventricular dilatation, aortic valve prolapse and aortic regurgitation and prevention of infective endocarditis [34] Open heart surgery entails closure directly (if small) or patch (Gore-tex, Dacron, bovine or autologous pericardium). (figure 6)

Patent ductus arteriosus (PDA) is the persistence of the fetal ductus arteriosus beyond two months after birth. This results in increased left heart return and failure to thrive with congestive cardiac failure. Most PDA's are amenable to closure by device (intervention) and surgery is done infrequently. Surgical approach is via left posterolateral thoracotomy. (figure 7) [35]

Aortopulmonary window (APW) is a communication between the aorta and pulmonary artery resulting in a large left-to-right shunt. This condition requires early surgical closure. This condition is rare and requires early surgery as there is steal from the systemic circulation. Surgical approaches vary but basically consist of division of the APW with suturing of a patch [36]

Double outlet right ventricle is a separate and complex entity wherein both great vessels arise predominantly from the right ventricle. When there is a VSD, this results in unrestricted pulmonary blood flow and physiologically is a left-to-right shunt. The classification of DORV by Lev is self-explanatory and the varying anatomy decides the clinical presentation which varies from simple left-to-right shunt, tetralogy of Fallot physiology, transposition of great

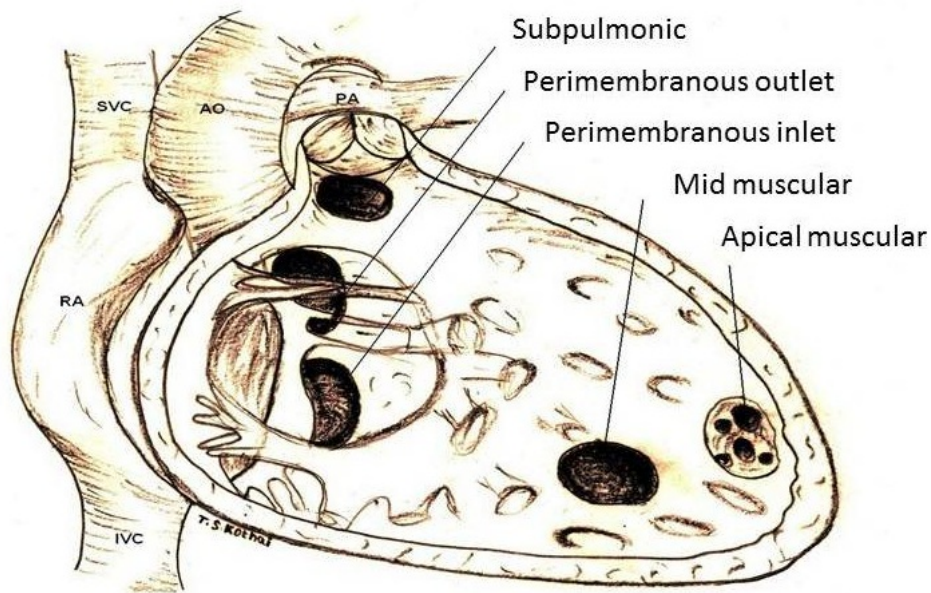


Figure 6. Ventricular Septal Defects.

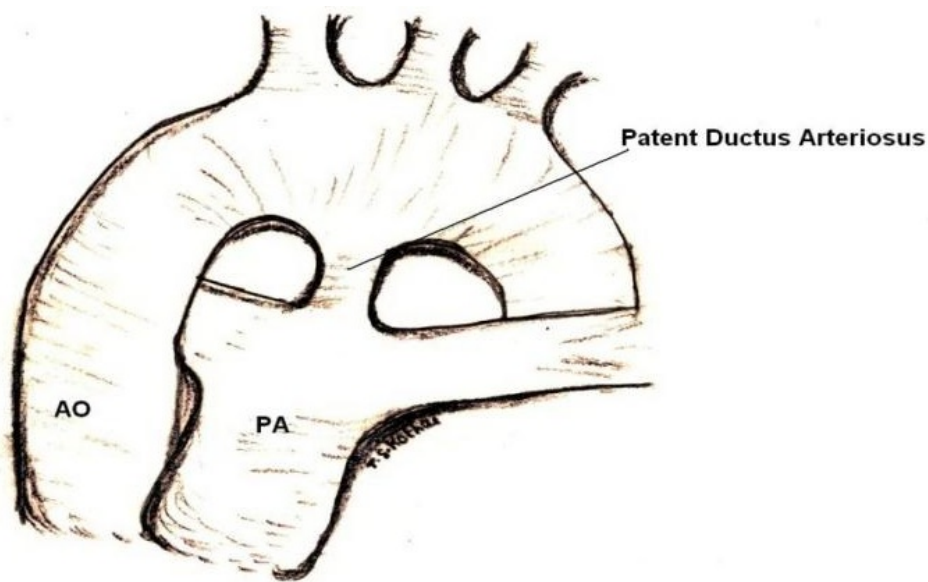


Figure 7. Patent Ductus Arteriosus

arteries physiology to single ventricle physiology. They occur with VSD which then presents as increased pulmonary blood flow. The treatment is surgical closure of VSD with routing of LV to drain into the aorta across the VSD. When they present with VSD and PS, the repair consists of VSD closure and relief of RVOTO with or without conduit. DORV with subpulmonic VSD Requires arterial switch and VSD closure [37]

Truncus arteriosus (figure 8) is a condition wherein the pulmonary arteries arise from the aorta directly. This results in torrential pulmonary blood flow and warrants early surgery in the

neonatal period. The PA is detached from the aorta and reconnected to the right ventricle utilizing a conduit and closing of the VSD. Alternatives include direct connection of RV-PA utilizing neighbouring tissue. Mid-term and long-term results are determined by the fate of the RVOT- the presence of pulmonary regurgitation and the deterioration of the conduit. [38].

Coarctation of aorta is narrowing of the aorta near the insertion of the ductus arteriosus to the descending aorta. This presents at various stages. Presentation in the neonatal period is usually as an emergency with closure of the ductus and sudden cessation of blood flow to the lower body. Treatment includes commencing prostaglandin to reopen the ductus, commencement of inotropes and correction of acidosis. Emergency surgical repair of coarctation is warranted. Surgical techniques are varied and each has merits and demerits. The preferred technique is resection of the coarctation and end-to-end anastomosis [39]. Alternatives include subclavian flap plasty and patch plasty. Interruption is total disconnection of the aorta and is usually associated with VSD. The classification is based on the location of the interruption (type A is distal to the left subclavian artery, type B is distal arch and type C proximal arch). Surgery is complex and involves disconnection of the PDA which supplies blood to the lower body and reconnection of the two ends of the aorta. The VSD is closed [40].

Pulmonary vein anomalies are rare and can present as stenosis of individual veins or as they enter the LA as a confluence. Prognosis is poor when all four pulmonary veins are involved [41]. Surgery or balloon dilatation are both associated with high rates of restenosis.

18.2. Cyanotic congenital cardiac conditions

The congenital cardiac conditions are Tetralogy of Fallot (TOF), Transposition of Great Arteries (TGA), Total anomalous pulmonary venous connection (TAPVC), Tricuspid atresia (TA) and Truncus arteriosus.

Tetralogy of Fallot (figure 8) is the commonest cyanotic congenital cardiac condition. The condition described originally consists of four components- VSD, overriding of aorta, right ventricular outflow tract obstruction (RVOTO) and right ventricular hypertrophy. The extent and severity of RVOTO accounts for the variation in symptoms. Babies present early with cyanosis or cyanotic spells (due to infundibular spasm). Surgical strategies vary in different parts of the world. In the neonatal period, whenever the baby presents with symptoms, the tendency is surgical correction with closure of VSD and judicious relief of RVOTO. However, most centers follow the policy of performing Blalock-Taussig shunt if the baby presents with symptoms at less than 6 months of age or less than 5 kgs. If older, surgical correction is attempted [42]. B-T shunt is still performed if the branch pulmonary arteries are hypoplastic or there are significant co-morbidities that preclude placing the baby on cardiopulmonary bypass. The postoperative course varies depending on the anatomical variations. Right heart failure is common due to the non-compliant RV and diastolic dysfunction.

B-T shunt is the connection of an artificial conduit between a branch of the aorta and one of the pulmonary arteries. This is a palliative surgery and is associated with complications such as blocked shunt precipitating cardiac arrest, low cardiac output, and pulmonary artery distortion. Historically, there have been several systemic-pulmonary artery shunts. These have

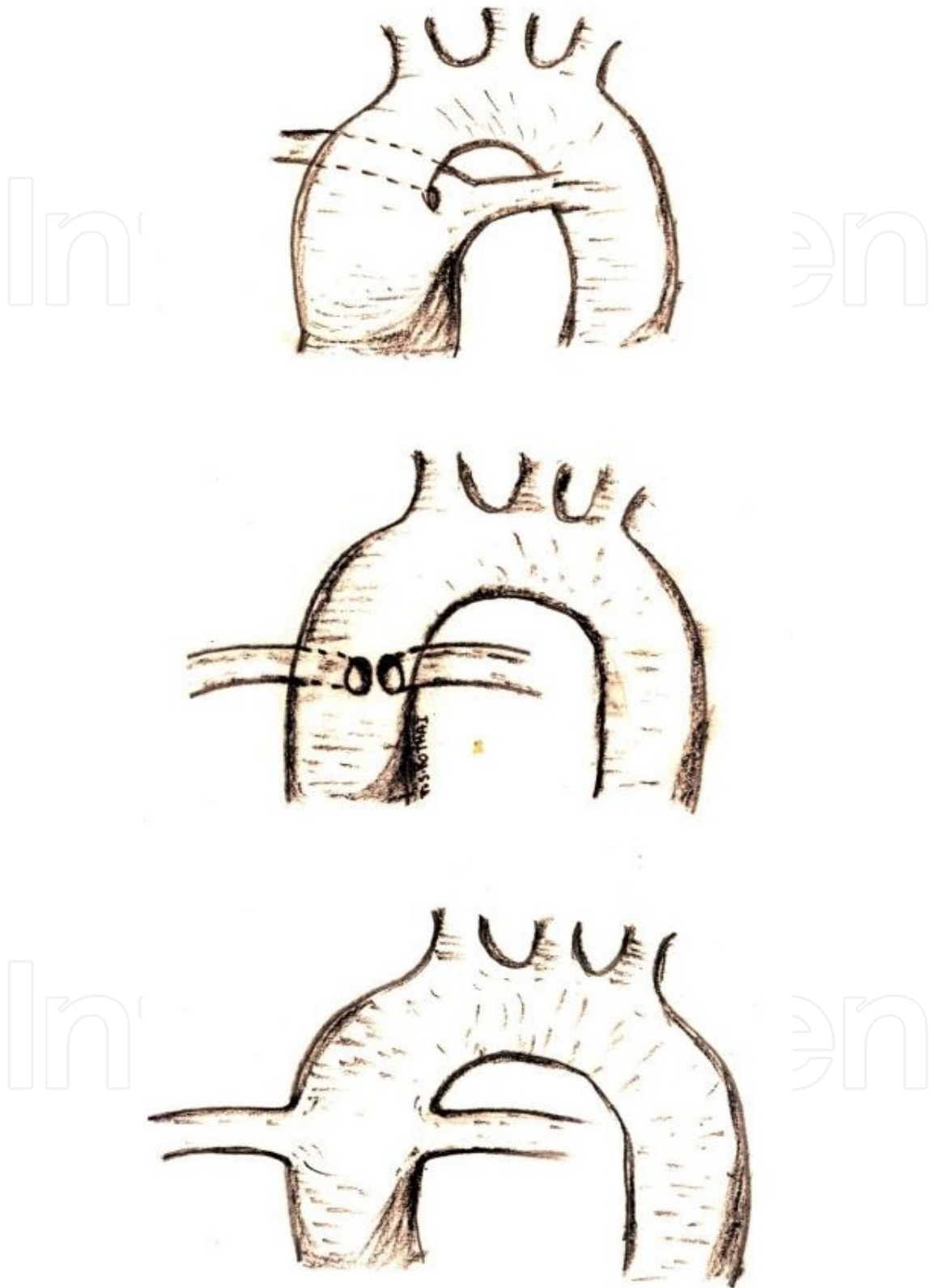


Figure 8. Types of Truncus Arteriosus

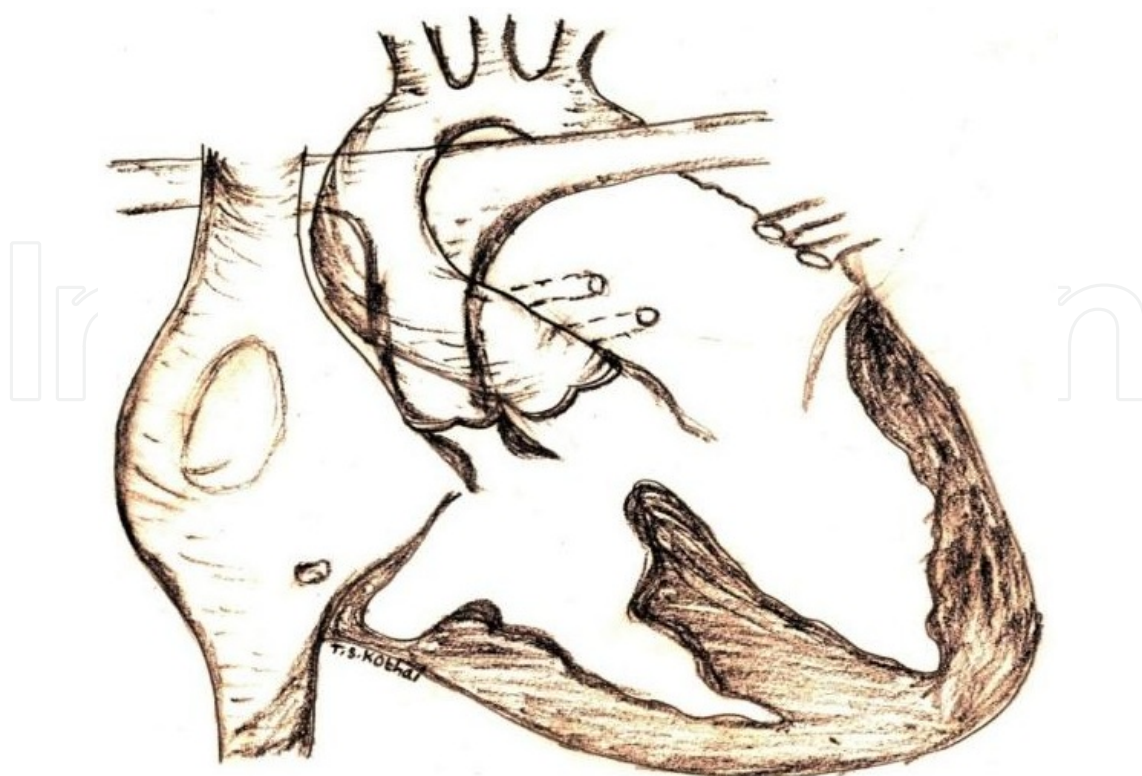


Figure 9. Tetralogy of Fallot showing a) VSD b) over ride of aorta c) Right ventricular hypertrophy and d) right ventricular outflow obstruction.

been anastomosis between ascending aorta and RPA (Waterston's) and descending aorta and LPA (Pott's). These shunts have resulted in uncontrolled pulmonary blood flow and gross distortion of pulmonary arteries.

Repair of TOF carries excellent results- less than 5% mortality in most centers. Potential complications are right heart failure, residual VSD, residual RVOTO and complete heart block. RVOTO relief varies from simple removal of obstructing muscle bundles to subannular patch to transannular patch. Transannular patch entails incising across the pulmonary valve annulus and results in free pulmonary regurgitation. This can cause right heart failure both acutely and in the long-term. Late pulmonary valve replacement is common in TOF repairs 10-15 years postoperatively. Surgeons are prophylactically placing bicuspid valves in the pulmonary position at the first surgery. [43] This improves immediate outcomes and as well as the long-term results.

Transposition of great arteries is the second commonest cyanotic condition. This presents in the neonatal period with the aorta arising from the right ventricle and pulmonary artery from the left ventricle. 50% are born with intact ventricular septum, 25% with VSD and 25% with VSD and PS. Senning's and Mustard's are procedures of historical importance. These are atrial switch procedures wherein the right atrium is directed to drain into the left ventricle and left atrium is directed to right ventricle. The disadvantage of this procedure is that the right

ventricle is placed in the systemic circulation. The RV is not designed to sustain the systemic circulation for long periods and many of these patients develop heart failure later and are candidates for heart transplant. Arterial switch operation is the transfer of coronary arteries from aorta to the pulmonary arteries. This results in switching the left ventricle to the systemic circulation.

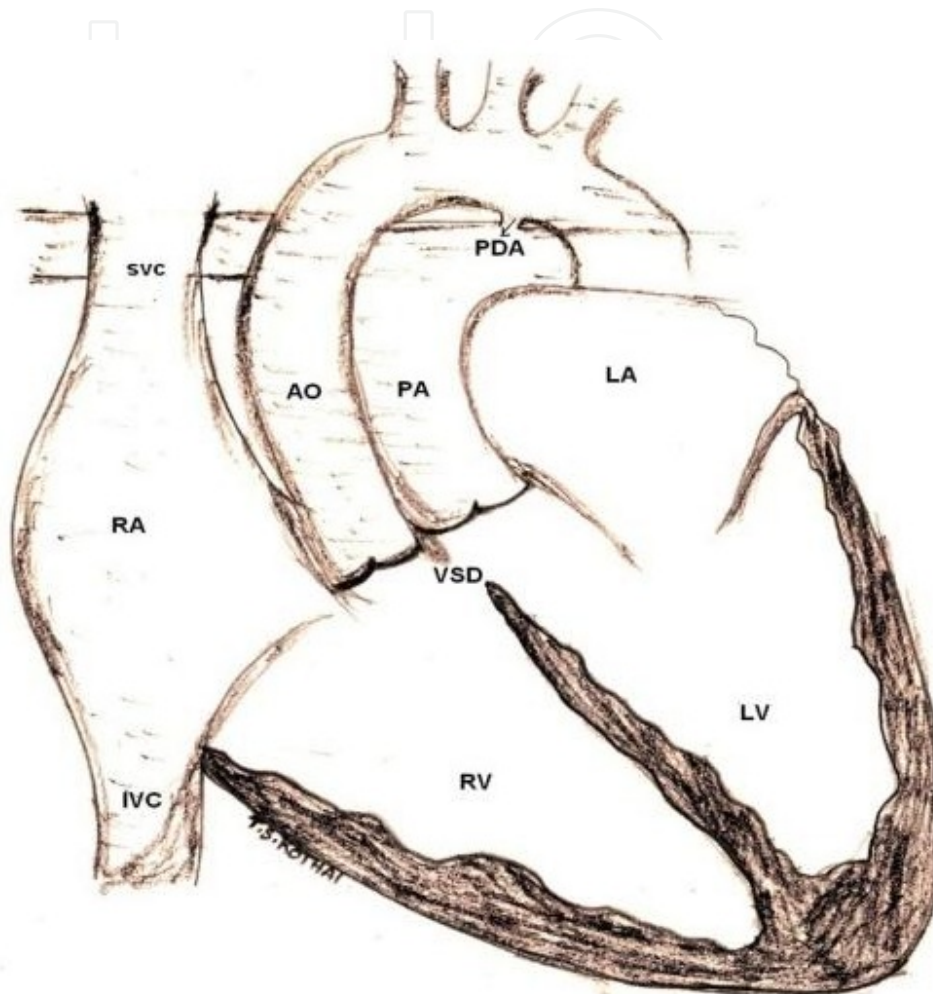


Figure 10. Transposition of Great Arteries

Babies with TGA and intact ventricular septum should ideally be operated within 2-3 weeks. The left ventricle loses its ability to sustain the systemic circulation beyond this as it adapts to the lower pulmonary artery pressures. When these patients present late, the options are to proceed with atrial switch which has lesser mortality and rapid two-stage arterial switch. The latter procedure is a staged procedure wherein the PA is banded and B-T shunt is created [44]. This creates the pressure and volume overload on the LV thereby retraining the LV to accept the systemic load. This is a relatively high-risk procedure. Serial echocardiographic assessment aids the surgeon to time the arterial switch once the LV is retrained.

Arterial switch is a worthwhile operation as the long-term results are excellent. Immediate postoperative complications are low cardiac output and myocardial ischemia due to coronary

insufficiency. Long-term complications are neoaortic regurgitation, neo-suprapulmonary stenosis and coronary ischemia [45] These occur in less than 10% of patients.

Total pulmonary venous connection (TAPVC) is a common congenital cardiac condition. Darling's classification divides TAPVC into 4 types- supracardiac, cardiac, infracardiac and mixed. The basic issue is that the pulmonary veins are not attached to the left atrium, form a common pulmonary venous chamber (CPVC) and drain via a communicating vein to some component of the systemic circulation. The commonest is supracardiac- the left vertical vein arising from the CPVC drains to the left innominate vein or less commonly, to the SVC just cranial to the SVC-RA junction. The cardiac variant usually joins the coronary sinus. The infracardiac type has a descending vertical vein draining vertically across the diaphragm to the portal vein or IVC. Mixed types are varying combinations of these. The presentation depends on whether there is obstruction in the pulmonary venous pathway or not. In supracardiac, the vertical vein may obstruct near the innominate vein or at the level of the ASD- this has to be completely unrestrictive or the child will present with obstructive pulmonary venous symptoms such as breathlessness, and even pulmonary edema. Cardiac type can have obstruction where the CPVC joins the coronary sinus or at the ASD. The infracardiac is the commonest type to present with obstruction as blood has to pass through the liver. This is one of the commonest neonatal cardiac emergencies. Surgery varies depending on the anatomy. Supracardiac TAPVC can be addressed by surgery - creation of anastomosis at the back of the heart between the Left atrium and CPVC. The creation of the anastomosis is dependent on the surgeon's preference- either working through the ASD, lifting the heart to the right, working lateral to the RA, working between the SVC and aorta or Shumacker's repair. Cardiac TAPVC is relatively simple- the coronary sinus is unroofed and pulmonary veins are committed to the LA after partitioning of the atria. The infracardiac TAPVC is again by creation of anastomosis between the LA and CPVC with disconnection of the descending vertical vein.

Immediate postoperative complications are low cardiac output, pulmonary hypertensive crisis and pulmonary vein restenosis- either early or late, at the anastomosis or in the individual pulmonary veins.

Tricuspid atresia is the absence of the tricuspid valve. ASD is mandatory for survival. They can present unrestricted pulmonary blood flow (PBF), in which case they need pulmonary artery band, followed by Glenn followed by Fontan. If the neonate presents with decreased PBF, the first operation is a B-T shunt followed by Glenn and Fontan. At each stage, before the Glenn and Fontan operations, the baby undergoes cardiac catheterization and angiography. The Glenn operation consists of dividing SVC and suturing this to the RPA. This unloads the single ventricle. The Fontan operation consists of a conduit being placed between the IVC and RPA. The single ventricle is thus unloaded of the pulmonary circulation and pumps blood to two circulations in series instead of two ventricles pumping blood to two circulations in parallel. Low cardiac output may occur and long-term complications include protein-losing enteropathy, plastic bronchitis and cardiac failure.

There are variations in univentricular physiology. The treatment protocol is similar to the one followed for tricuspid atresia. The common variant of this is hypoplastic left heart syndrome (HLHS) wherein the aorta is hypoplastic and a complex operation called Norwood is per-

formed in the neonatal period. The PA is disconnected and reconnected to the systemic circulation to enhance the arch. The pulmonary circulation is provided by a B-T shunt. The patient subsequently undergoes Glenn and Fontan procedures.

Corrected transposition of great arteries (CCTGA) or l-TGA consists of atrio-ventricular and ventriculo-arterial discordance. The child may present with intact ventricular septum. The disadvantage lies in the fact that the RV is in the systemic circulation and will fail over a period of time. The child may present with VSD and with or without pulmonary stenosis. There are two approaches- anatomic and physiologic repairs. The physiologic approach consists of addressing the various lesions- VSD and PS thereby leaving the RV in the systemic circulation with doubtful long-term outcomes. The anatomic repair consists of double switch. The procedure consists of performing atrial switch and arterial switch. This eventually results in the LV being in the systemic circulation. If there is a VSD, this is surgically closed. The chances of complete heart block requiring a pacemaker is high as the A-V node and Bundle of His are in an abnormal position. If there is a PS, the procedure is an atrial switch-Rastelli- the RV is connected to the PA via conduit. These are complex procedures carrying significant morbidity and mortality [46] These children present with left A-V valve regurgitation as well. This is the tricuspid valve functioning as the systemic valve. A simplified approach to this condition is to perform Glenn followed by Fontan procedure if they present with VSD with significant PS. This carries lesser mortality than the above procedures. However, the Fontan circulation is an imperfect state and has its own disadvantages.

18.3. Congenital lesions of valves, aorta and coronary arteries

The four valves are prone to various congenital anomalies:

1. Tricuspid valve- Ebstein's anomaly is the commonest congenital anomaly of the TV. There is apical displacement of the septal and postero-inferior leaflets with a sail-like anterior leaflet. This results in severe TR. The RA is dilated with RV dilatation and dysfunction. This condition is associated with accessory conduction pathways which predispose to supraventricular arrhythmias. This is diagnosed from the presence of delta waves in the ECG. Criteria for surgery are presence of dyspnoea NYHA III-IV, cardiomegaly (CTR ratio > 0.65) and cyanosis in the presence of ASD. LV dysfunction is noted when cases present late due to RV dysfunction causing LV to be affected by the phenomenon-of ventricular interdependence. Neonatal Ebstein's is a particular difficult subset to treat and the Starne's operation has been described- elective closure of the tricuspid valve and B-T shunt followed by staging to Glenn and Fontan.(figure 10).

Children and adults with Ebstein's anomaly require tricuspid valve repair. There are various techniques described such as Danielson's [47], Carpentier's, Cone [48] and Stanford [49] technique and variations on these with proponents for each. The repair aims to achieve tricuspid competence, without compromising RV cavity if possible. Some techniques aim to obliterate the atrialised RV and others ignore it. Complications include severe RV dysfunction, arrhythmias, low cardiac output. Glenn is performed by the surgeon if he feels the RV will not be able to cope. If tricuspid regurgitation is significant, the tricuspid valve is electively

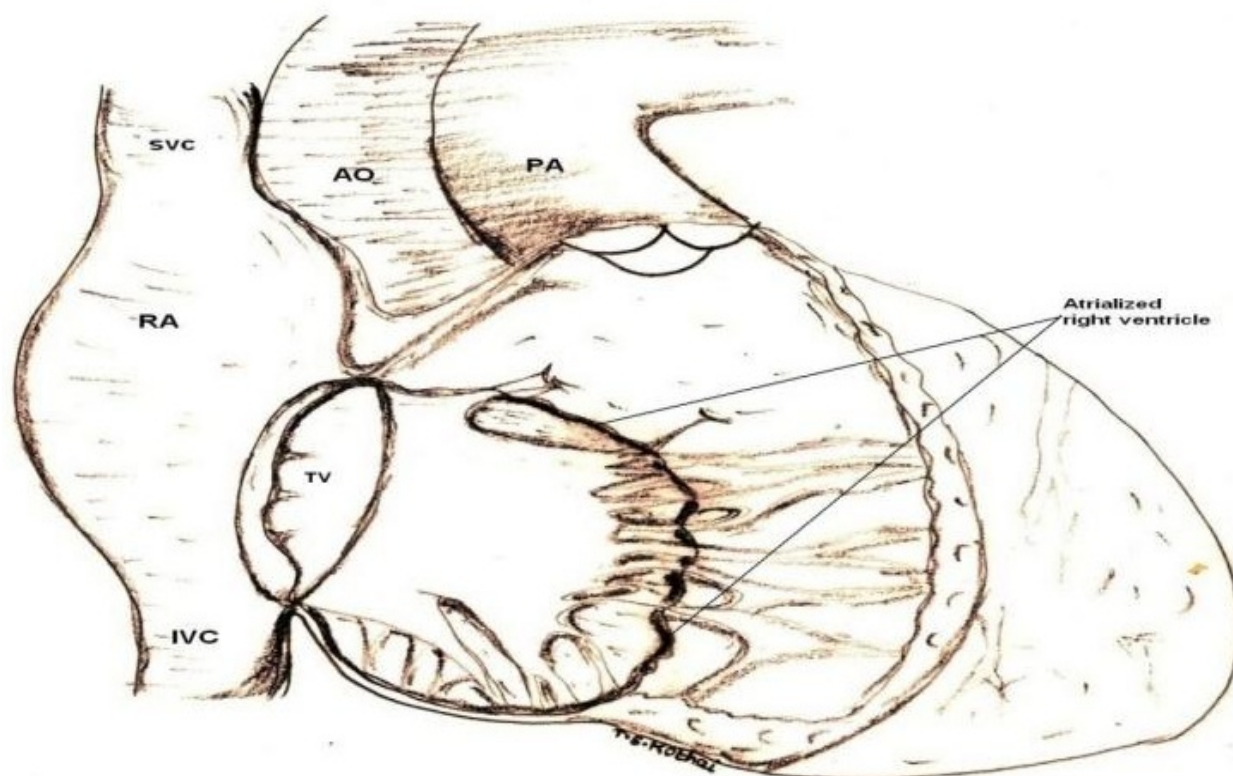


Figure 11. Ebstein's anomaly of tricuspid valve showing atrialised RV and displaced tricuspid leaflets.

replaced. Bioprosthetic valves are preferred in the tricuspid position as the circulation is sluggish compared to the left heart and mechanical valves in this position are prone to clotting and there have been reports of sudden death. Bioprosthetic valves require reoperation as they deteriorate over a period but reoperations are not associated with increased mortality.

Other causes of congenital TR are due congenital dysplastic leaflets. Various standard valve repair strategies are employed such as artificial Chordae, closure of clefts and commissures, and leaflet enhancements with commissural annuloplasty or ring annuloplasty.

2. Pulmonary valve- Congenital anomalies of the PV present as pulmonary stenosis. Intervention is indicated when the gradient > 50 mm Hg. This is usually performed by the interventional cardiologist. Most cases are amenable to balloon valvuloplasty. Surgery is only indicated if there is supra-annular narrowing, annular hypoplasia or dysplastic leaflets. Surgical relief is by open pulmonary valvotomy with or without transannular patch. Patients can present with isolated infundibular obstruction. This requires open heart surgery- either resection of RVOT muscle bundles and if necessary, subannular patch.

When patients present with free pulmonary regurgitation following TOF repair, reoperation is indicated if children are symptomatic, the RV dilates or the patient presents with ventricular tachycardia. Surgical options include pulmonary valve replacement with pulmonary or aortic

homografts, bovine jugular vein conduits and bioprosthetic valves. These valves are of limited durability and will require re-replacement. Percutaneous deployment of pulmonary valves has clinically employed with limited success.

2. Mitral valve- Congenital anomalies MV can be either mitral stenosis (MS) or mitral regurgitation (MR). Variants of MS include commissural fusion, parachute MV (single papillary muscle), hammock MV and variations in anomalies of the annulus, leaflets, chordae and papillary muscles. Congenital MR is due to the following mechanisms: Carpentier's classification- type 1-normal leaflets, type 2-leaflet prolapsed, type 3-leaflet restriction. If there is perforation in the leaflet, this is closed with a patch. If the annulus is dilated, this is reduced either by suture annuloplasty, commissural annuloplasty or ring annuloplasty. In leaflet prolapsed, the use of artificial chordate is gaining popularity. Contrary to the belief that this is contraindicated in children as they may outgrow their chordae, this has not been found to be the case as corresponding growth of the papillary muscle compensates. Chordal transposition or chordal shortening are described with good results. Leaflet restriction is addressed by releasing tethering secondary and tertiary chordae and leaflet enhancement with pericardial patch. Repair is always preferable in children at the expense of residual MS/MR as mitral valve replacement carries high risk in children due to their small annulus. Bioprosthetic valves degenerate rapidly due to the accelerated calcium metabolism of adolescence and mechanical valves require anticoagulation which is difficult to manage in children. Small valves are quickly outgrown by the child.
3. Aortic valve- Commonest AV lesion is congenital bicuspid aortic valve. This usually presents with stenosis. The optimal treatment when obstruction is significant is balloon valvuloplasty. This is associated with greater recurrence but delaying surgery is always preferable till the child is bigger and the annulus is larger. Congenital AS may present as unicuspid valve as well. Open aortic valvotomy is a simple procedure wherein the surgeon splits the valve judiciously at the commissures. Bicuspid valve can present with AR as well. Repair is still preferable and various valve preservation techniques are described. Residual AR is preferable to aortic valve replacement in infants and children. AVR in children varies from mechanical valve replacement in older children, Ross procedure wherein the patient's PV is placed in the aortic position and conduit placed in the pulmonary position and aortic homograft root replacement. Anticoagulation is not required for Ross and homografts with the former carrying the advantage of increased durability of the aortic autograft. Mechanical valves, particularly if small, are fraught with problems particularly outgrowth.

Children, particularly those with Marfan's and other connective tissue disorder, can present with aneurysmal dilatation of the aorta. The indications for surgery are similar to those for adults with the aim being to prevent rupture. Surgical options include replacement of the ascending aorta with an interposition graft and Bentall procedure wherein the whole aortic root is replaced with composite graft including a mechanical valve and reimplantation of the coronary arteries if there is associated AR. Current concepts are to preserve the native valve

whenever possible and hence, further surgical options are to replace ascending aorta and repair the AV or root-preserving surgery.

Coronary artery anomalies are uncommon and include anomalous origin of the left coronary artery from pulmonary artery (ALCAPA), coronary arterio-venous fistula and coronary artery aneurysms. ALCAPA characteristically presents with severe LV dysfunction and mitral regurgitation. The problems are two-fold: connection of the left coronary artery to the PA which results in myocardial ischemia and the left-to-right shunt ensuing. Ideal procedure is urgent surgery and re-implantation of the left coronary to the aorta. Takeuchi repair or its modifications to baffle the left coronary to the aorta has also been described. Less than ideal is to tie off the left main and leave the patient on a single coronary. Adults have been managed by ligation of left main and coronary artery bypass grafting.

18.4. Congenital tracheal anomalies and vascular rings

These are rare conditions. Congenital tracheal stenosis consists of localised or long-segment stenosis of trachea due to presence of complete tracheal rings. These are associated with cardiac lesions such as TOF. The patients present with stridor. Surgery is the treatment of choice. These include slide plasty, resection and end-to-end anastomosis, homograft replacement and patch plasty of trachea simultaneous with the cardiac repair.

The two common vascular anomalies that present are double aortic arch and left pulmonary artery sling. Double aortic arch [50] is due to the persistence of both right and left dorsal aortic arches. Symptoms are related to the compression of the trachea and esophagus by the relevant vascular structures. Surgery for double arch is via left thoracotomy, division of the PDA/ligamentum arteriosum and surgical division of the smaller arch distal to the corresponding subclavian artery. LPA sling is associated in 50-65% of cases with tracheal stenosis due to complete rings. Surgery is by reimplantation of the LPA onto the MPA either via left thoracotomy or median sternotomy on cardiopulmonary bypass [51]

18.5. Adult congenital cardiac surgery

Children who grow into adults with congenital heart disease come under this category.

Left-to-right shunts: Atrial septal defect of all types can present in adulthood and are usually operable. A small percentage present with irreversible pulmonary hypertension. Operations on adults provide a survival benefit upto 25 years of age, but beyond that, the main indication is improving quality of life, prevention of paradoxical embolism and most importantly, prevent the onset of atrial fibrillation.

Ventricular septal defect rarely present in adulthood as they are operable only as children. The only odd case maybe the rare ones with large VSD that have not become Eisenmenger's and the one who present with aortic valve prolapse or along with aortic regurgitation and/ or ruptured sinus of Valsalva aneurysm. The latter characteristically present in middle age. The other common subset is those adults who present with VSD and RVOT obstruction (double chamber right ventricle or adults with closing VSD and acquired RVOT obstruction-gazzulization.)

Adults do occasionally present with PDA- the large ones are rarely still operable and the small ones can be occluded by the interventional cardiologist. Surgery in PDA with pulmonary hypertension is high-risk as they are fragile and prone to catastrophic tears during surgery- they may be calcified. Surgery is more complex- requiring cardiopulmonary bypass.

Coarctation is not uncommon in presentation as an adult, either as a primary coarctation or re-coarctation. Surgery is high-risk, particularly with tight coarctation with multiple collaterals. Entry into the chest is associated with significant bleeding and very often, the coarct segment has to be excised and replaced with an interposition graft. The aortic tissue is usually friable and is prone to tear. A bypass graft from the left subclavian artery to the descending aorta may then be preferable. Recoarctation is better dealt with balloon plasty and stenting. Adults with coarctation may be managed with covered stents.

Tetralogy of Fallot may present in adulthood. They are amenable to corrective surgery provided criteria are met such as adequacy of pulmonary arteries and there are no significant collaterals. They may need preoperative embolization of collaterals and intraoperative RV-PA conduit as they may not tolerate free pulmonary regurgitation in the event of transannular patch. The children who underwent repair as children may present in adulthood with RV dysfunction due to free PR. They require redo sternotomy and insertion of RV-PA conduit. A subset of these patients presents as adults with aortic regurgitation and/or ascending aortic aneurysm and require AVR with or without ascending aorta replacement.

Patients may present de novo with RVOT obstruction at various levels. These are amenable to surgery or intervention based on various criteria.

Children who underwent Senning's or Mustard's procedure from transposition of great arteries (TGA) present in adulthood with residual defects such as baffle leaks or with cardiac failure as the RV in the systemic position is prone to failure. These patients require heart transplant or conversion to arterial switch following PA banding to retrain the LV. These are high-risk procedures.

Children with single ventricle physiology present in adulthood requiring Fontan procedure. Some have had atriopulmonary Fontan and require conversion to an extracardiac Fontan if the former surgery fails.

Ebstein's anomaly, cor triatriatum sinister (membrane in the left atrium obstructing pulmonary veins), ALCAPA and TAPVC with unrestricted ASD are conditions with which patients may present as adults for surgery. Congenital corrected transposition of great arteries, with VSD and PS may also present late.

Children who have undergone arterial switch as infants present as adults with coronary issues, neo-pulmonarysupravalvular stenosis, and neo-aortic regurgitation. These patients require stenting of their coronary artery obstructions, coronary artery bypass grafting, patch plasty of supravalvular obstruction and replacement of the aortic valve.

19. Arrhythmias, pacemakers and defibrillators

The congenital population is prone to postoperative complete heart block and even nodal arrhythmias are not tolerated by the population with single ventricle physiology. These children will require to be under the care of the electrophysiologist. They will need lifelong pacemaker changes and lead changes. Arrhythmias are common such as atrial fibrillation and those related to accessory bundle pathways. Children with dilated RV/LV are prone to malignant ventricular arrhythmias such as ventricular tachycardia and ventricular fibrillation. These require monitoring and insertion of AICD (automatic internal cardioverter-defibrillator). Patients with chronic atrial fibrillation may benefit from the Maze procedure (antiarrhythmic surgery). This procedure may involve either right atrium or left atrium or both.

20. Future of congenital cardiac surgery

- 1. Rise of interventional cardiology:** The interventional cardiologists have taken over many of the procedures formerly done exclusively by the surgeons. These include atrial septal defect (ostium secundum), and patent ductus arteriosus closure. Certain VSD's are being closed by the cardiologists. Open pulmonary valvotomy is a procedure of historical importance and open aortic valvotomy is rarely undertaken as a primary procedure. Procedures which will be undertaken with increasing aggressiveness by the interventionalist include ductal stenting, and RVOT stenting as a substitute for B-T shunt, and percutaneous pulmonary valve replacement. Percutaneous maze procedures, percutaneous mitral valve repairs, and percutaneous completion Fontan are on the anvil. Lesions such as aortopulmonary window, aorto-cameral tunnel and coronary a-v fistulae are amenable to interventional strategies. This will result in a change in the spectrum of surgeries done- surgeries will become more complex and the number of simple surgeries will dwindle. Hybrid procedures will rise wherein the cardiologist and cardiac surgeon will work together.
- 2. Fetal echocardiography:** Antenatal diagnosis of congenital cardiac conditions is possible at 18-20 weeks of gestational age. This will empower parents with the option of medical termination of pregnancy. This will reduce the number of children requiring heart surgery.
- 3. Robotic cardiac surgery/ thoracoscopic surgery:** Certain operations may be performed by the surgeon sitting at the console or maybe even in another part of the world utilising computer-controlled robotic arms. Thoracoscopic surgery may be an alternative for PDA.
- 4. Disappearance of valve replacements:** Surgeons will become increasingly skilled at preserving the patient's own valve by mastering the skill of repairing rather than replacing with substandard alternatives.

5. **Genetic engineering:** Valves manufactured from the patient's own genetic material will demonstrate greater durability and biocompatibility and may become available off the shelf.
6. **Heart and heart-lung transplant:** There is a huge population of patients with Eisenmenger's syndrome who will eventually require transplant. Transplant restrictions such as availability of organs (genetically manufactured) and rejection will be overcome.
7. **Advancements in perfusion, anaesthesia and intensive care:** Better perfusion technology and techniques to avoid cardiopulmonary bypass with its deleterious effects on children will improve results of cardiac surgery. Pharmacological advances and newer drugs will provide for better outcomes via improved anaesthesia and postoperative strategies.

21. Quality improvement and risk stratification in congenital cardiac surgery

There are various risk-stratification scores in congenital cardiac surgery such as RACHS-1 and Aristotle Comprehensive Complexity Score. This places into perspective the mortality and morbidity rates of the hospital relative to the complexity of the cases they accept.

Databases exist such as the Society of Thoracic Surgeons database (STS) and EACTS congenital database which allows surgeons to compare the results of their surgery on various diagnoses with those of various others across the world.

22. Conclusion

Pediatric cardiac surgery is a noble pursuit, the healing of a sick heart by surgery in a fragile child. The future remains exciting with many anticipated developments. Many complex conditions will disappear as antenatal diagnosis becomes a norm and medical termination of pregnancy the rule. There will be an explosive growth in the number of adults presenting with congenital heart disease. This would present a huge stress on the healthcare providers and we must be prepared for this.

Surgeons will have to sharpen their skills as simple conditions will disappear from their operating list, which would be dealt with by interventional cardiologist. It would be replaced by complex conditions, adolescents and adults with congenital problems, requiring reoperations. They will have to accept fresh challenges- a surgeon cannot just replace a valve, but figure out strategies and surgical techniques to repair the patient's valve. The bar is thus raised.

In the late 1990's and early part of this century, mortality rate was the parameter by which a surgeon or unit was judged. This will be replaced by tougher parameters- complication rates and success rates with the more complex congenital cardiac conditions. Genetic engineering and transplant technology will have the greatest impact on the future of congenital cardiac

surgery. Emphasis will be on the quality of life of survivors and normalizing their lives as much as possible. The future is indeed exciting and full of wonderful possibilities!

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