Review Article

CONGENITAL CARDIAC DEFECTS: WHEN TO INTERVENE

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Delayed diagnosis and referral of Pediatric Cardiac patients in addition to inability of parents to financially undertake pediatric cardiac surgery have resulted in several patients becoming inoperable. Improved survival after early repair has also resulted in earlier age of surgery. Several procedures are now feasible in the neonatal period and in addition reparative procedures which required an addition palliative surgery are now performed in a single step. The objective of this article is to review contemporary recommendations for timing for surgery and interventions.

Key words: ASD, Cardiac Surgery, Neonate, Transposition, VSD.

INTRODUCTION

THE timing of surgery of any cardiac defect is an integral of a variety of factors that are related to the child's defect and those related to the outcomes that can be provided in a pediatric cardiac unit. Optimal results can best be obtained in dedicated pediatric units with high volume. To provide results parallel to those published from Centers of Excellence in developed countries, several variables have to be optimized: early accurate diagnosis, pre-operative stabilization (occasionally with interventions like balloon septostomy), intra-operative management, post-operative management. Once these are optimized, surgery performed at appropriate time can result in excellent surgical results with no long term issues in majority of cases. This chapter focuses on the timing for intervention, surgical and catheter based, in congenital cardiac lesions.

PRE-TRICUSPID SHUNTS

Atrial septal defects / partial anomalous Pulmonary venous return (ASD/PAPVR)

Atrial septal defects are usually operated at 2 years of age or anytime after that when they have been diagnosed. Very rarely (approximately 10%) an isolated atrial septal defect may require closure in infancy due to difficult to control CHF [1].

Contemporary surgical outcome of this lesion is 100%. Time related survival of patients operated for ASD or PAPVC during the first few years of life is that of the matched general population implying there is no late attrition of these patients and survival continues in the second decade and beyond. Sinus venosus ASD is the lesion in this group associated with late morbidity.

Features to follow carefully would be RV diastolic dimensions, (especially if the closure was preformed at an older age), atrial arrhythmia and in case of PAPVR, return of draining pulmonary segments. Those with atrial arrhythmias especially atrial fibrillation run the risk of thrombo-embolism and therefore need anticoagulation.

A recently published study looking at long-term outcome of sinus venosus ASD (SVASD) noted outcomes which require attention. The authors reviewed outcomes of 115 patients (mean age 34 years) with SVASD who had repair. Complete follow-up was available in 95% patients at 144+/-99 months. Symptomatic deterioration was noted in 17 patients (16%). At follow-up, 7 (6%) of 108 patients had sinus node dysfunction, a permanent pacemaker, or both, and 15 (14%) of 108 patients had atrial fibrillation. Older age at repair was predictive of postoperative atrial fibrillation. In conclusion a significant percent of patients required electrical management of the heart [2].

POST-TRICUSPID SHUNTS

Ventricular septal defects

Ventricular septal defects are the commonest cardiac lesion. Unfortunately this lesion is dealt with most inappropriately as regards timing for surgery resulting in irreversible pulmonary hypertension. Patients that are diagnosed in early infancy are most likely due to large unrestrictive defects. This subset will present with early onset, refractory congestive heart failure and severe failure

to thrive. Such lesions should be closed within the first 3-4 months of life or when diagnosed. Presence of coarctation or aortic interruption will require neonatal surgical intervention. Moderate sized defects need careful follow up and should be offered closure based on the merit of symptomatology. The timing most often should be within the first two years of life. Special circumstances that need careful serial echocardiographic evaluation for early surgery are development of progressive pulmonary hypertension, new onset aortic or tricuspid insufficiency, secondary infundibular stenosis (Gassul's effect) or an episode of infective endocarditis. Defects in anatomic locations that are less likely to close spontaneously (inlet defects, subpulmonary lesions) should be offered early surgery. Presence of Trisomy 21 enhances the risk for early development of pulmonary vascular disease and therefore need closure within the first 9 months.

Ventricular septal defect is one of the lesions where the long term outcome is excellent if no residual lesion has been left behind [3]. Premature late death occurs in less than 2.5% of patients provided the pulmonary resistance was preoperatively judged to be low. Patient who had high preoperative pulmonary vascular resistance may die from progressive pulmonary vascular disease. Thus it must be emphasized that closure during the first year of life is curative. If timely closure is done the physical growth becomes normal for weight, length and head circumference by age of one year.

PATENT DUCTUS ARTERIOSUS

Symptomatic lesions with failure to thrive should be closed irrespective of the age or weight of the child. Patients with milder symptoms can be timed for closure at around 3 months of age. Asymptomatic children may be offered closure to prevent possible infective endocarditis. Extremely low birth weight babies with PDA who are ventilator dependent should have PDA ligation (especially if failed/contraindicated pharmacological therapy) early before the onset of end organ dysfunction.

Life expectancy is normal after closure of an uncomplicated PDA in infancy or childhood. With older age at operation or preoperative high pulmonary vascular resistance late deaths may result from progression of pulmonary hypertension.

ATRIOVENTRICULAR SEPTAL DEFECTS (AVSD)

Elective surgical closure of the complete forms of AVSD should be done between 3-6 months of life or earlier if severe failure to thrive / CHF. Partial and transitional forms of this lesion will need closure within the first two years of life. Early intervention is required in the subset of patients who have severe mitral incompetence or left ventricular outflow tract obstruction.

Most long term survivors (88%) are in excellent NYHA status. These defects once repaired and are beyond the early hazard phase (which is steep for the initial 6-9 months after surgery) show a very small but appreciable constant hazard phase. This means that these patients need regular follow-up even in the long term.

The main reasons for concern are left AV valve dysfunction (regurgitation/stenosis), left ventricular outflow tract obstruction (more after a partial AV septal defect repair than complete), residual pulmonary hypertension, and rarely complete heart blocks or supra ventricular arrhythmias. It should be noted that presence of Trisomy 21 does not adversely effect the outcome of these defects. Incidentally a study published in 1995 with follow up of 203 patients concluded that mortality has decreased over the decades from 19% before 1980 to 3% after 1990. The 10 year survival is 90% and all patients are in NYHA Class I or II. Late reoperation was required in only 8/203 patients and their results indicate that complete atrioventricular septal defects can be repaired with low mortality and good intermediate to long-term results [4].

COARCTATION OF AORTA (CoA)

CoA presenting in the neonatal period is a duct dependent lesion requiring initial stabilization with prostaglandin E1 followed by neonatal repair. Neonates presenting with renal and mesenteric dysfunction (ductal shock) and those with severe LV dysfunction may require urgent salvage angioplasty followed by surgical repair. CoA presenting later should preferably be intervened within the first year of life or when diagnosed. In older children with borderline resting gradients across the CoA, a stress test may unmask an abnormal blood pressure response in the brachio-cephalic arteries.

In isolated coarctation of aorta long term survival is excellent. Long term issues include resting and exercise induced hypertension. Recurrence of coarctation has to be sought for actively. Commonly associated bicuspid aortic valve may show signs of stenosis by the 2nd or 3rd decade of life and may also be a source to infective endocarditis. New or late onset subaortic stenosis is also an issue to be looked for. If patch aortoplasty was performed care should be taken to evaluate for aneurysm formation in the long run. Cerebrovascular accidents are more common in patients with persistant hypertension and in those with Berry aneurysms.

In a late follow up of upto 50 years after coarctation repair, surprisingly 18% (45/274) patients died at a mean age of 34 years. Predictors of survival were age at operation and blood pressure at the first postoperative visit. The

authors noted long-term survival is significantly affected by age at operation, with the lowest mortality rates observed in patients who underwent surgery between 1 and 5 years of age. More than one-third of the survivors developed significant late cardiovascular abnormalities [5].

CYANOTIC HEART DISEASE

Truncus arterisus/aorto-pulmonary window

These cono-truncal septal defects should undergo repair within the neonatal period. Some patients who present later should be operated as early as possible after careful assessment of pulmonary vascular resistance.

Since a valved conduit is used for repair during early infancy, issues related to the valve necessitate reoperation or intervention or both. Pulmonary vascular disease often develops and progresses in older infants and children who did not undergo repair and therefore adversely affect survival in the long run. A recent single center review of 16 cases on intermediate term follow up indicate conduit and valve failure at mean follow up of 2.5 years in 67% of the patients [6].

Tetralogy of Fallot (TOF)

TOF with pulmonary stenosis that presents in very early infancy with cyanotic spell may undergo a palliative BT shunt. Patients presenting without a history of spell should be operated between 6-8 months of life or earlier if saturations are approaching 75%.

TOF with pulmonary atresia presenting in the newborn period with cyanosis is most likely a duct dependent lesion and hence will require stabilization with prostaglandin E1 followed by a palliative BT shunt. Other variables requiring objective assessment are the sources of pulmonary blood flow, degree of cyanosis or congestive heart failure and size and continuity of central pulmonary arteries. Based on the above stage unifocalization can be performed within the first year of life. In all cases of TOF with pulmonary atresia, complete repair is aimed at the age of 18-24 months.

TOF with pulmonary stenosis a heterogeneous group of patients that behave in a variety of ways in long term follow up. To answer the difficult question regarding "Surgical Cure" of Tetralogy we take lessons learnt from a large meta-analysis. This infers that time related survival of most patients after repair of TOF with pulmonary stenosis under proper circumstances is excellent, approaching that of general population, but the risk of death throughout life in "slightly greater" than general population.

During follow up of these patients important points to be noted are RV function, RV outflow tract aneurysms, pulmonary insufficiency (specially if a transannular patch has been placed during the repair) residual right ventricular outflow tract (RVOT) obstruction and recurrent / residual VSDs, late aortic insufficiency, tricuspid valve competence and ventricular or supra ventricular arrhythmias.

Progressive RV dilatation and dysfunction (preferably documented by MRI) should be sought for early. An ECG with QRS duration >180 ms puts these patients on a higher risk for sudden death from ventricular arrhythmias.

A recent study evaluated 95 patients with TOF on long term follow up for their RV function. Despite a large number of reoperations, this cohort of patients remained well with low incidence of sudden death with normal good RV and LV function. Aggressive intervention for right-sided hemodynamic abnormalities may have contributed to this outcome. Preserved ventricular function may herald a favorable long-term outlook in this group, the authors concluded [7].

In another multi-center study, 793 patients with Tetralogy were reviewed for late arrhythmia complications. Thirty-three patients developed sustained monomorphic ventricular tachycardia, 16 died suddenly, and 29 had new-onset sustained atrial flutter or fibrillation. Pulmonary regurgitation was the main underlying hemodynamic lesion for patients with ventricular tachycardia and sudden death, whereas tricuspid regurgitation was for those with atrial flutter/fibrillation [8].

THE SINGLE VENTRICLE

This terminology comprises a wide variety of diagnosis. These patients have to be placed on what in termed as a single ventricle or FONTAN palliation protocol. Patients with effective single ventricle have varied presentations. A large group presents with severe cyanosis and will need a palliative shunt preferably in the newborn period. Second subgroup presents with congestive heart failure and very mild or imperceptible cyanosis and will require neonatal pulmonary artery banding. The third subgroup presents with neonatal systemic shock and require initial stabilization with prostaglandin E1 followed by construction of an unobstructed systemic outflow path and a BT shunt. All these groups of palliated patients then will require a bidirectional GLENN (Superior cavo-pulmonary anastomosis) at the age of 6 months. The final surgical palliation involves routing the inferior vena caval blood into the pulmonary arteries called the FONTAN operation which is performed between 2-4 years of age.

The long term survival issues cannot be simplified because of the heterogeneity of the primary diagnosis for which a FONTAN pathway was offered. With the latest modifications in the staged palliation towards FONTAN operation mid term survival is approaching and exceeding

90%. There is present the late slow rising phase of the hazard curve. One modification is to fenestrate the baffle and this leads to some degree of acceptable cyanosis.

During the follow up important facts to review and expect are development of supra-ventricular tachy-arrhythmias, ventricular dysfunction, protein losing entero-pathy, serous cavity effusions, thromboembolic and neuro-logical complication (specially if a fenestrated FONTAN is done). Development of AV valve regurgitation is detri-mental and needs to be evaluated. Systemic hypertension and semi lunar valve incompetence promote load on the ventricle and need prompt attention. Obstruction to the FONTAN pathway and baffle leaks should also be looked into. These adolescents self learn to cope up with mild exercise intolerance.

In a recent review of 225 patients of double inlet single left ventricle (DILV) who underwent FONTAN operation in Mayo Clinic between 1974 to 2001, median age at operation was 9 years and median follow up was 12 years. The operative mortality after 1989 has decreased to 3%. Current health as described by the patients was good or excellent by 84% of patients, fair by 18%, and poor by 12%. The authors concluded that, the FONTAN operation for DILV is now performed with a low operative mortality rate. Long-term survival has improved, and most patients have good functional status [9].

TRANSPOSITION OF GREAT ARTERIES (TGA)

TGA can be broadly classified as simple transposition, TGA with VSD and TGA with VSD with PS. Simple transposition will require immediate stabilization with prostaglandin E1 followed by balloon atrial septostomy (if needed) and arterial switch operation within first 4 weeks of life. Later presentations will require retraining of the left ventricle in the form of a PA band and BT shunt. Presentations beyond the 3 months of life may be offered atrial switch operation (Senning or Mustard procedure). TGA with VSD should be corrected within 6 months. TGA with VSD with PS may require a palliative BT shunt in the early infancy if the saturations are low and later by the age of 18-24 months should be completely corrected by a Rastelli operation.

Complete transposition of the great arteries is a relatively common anomaly, which comprises 5 to 7% of all instances of cardiac malformations. Given the decreasing mortality rates associated with a neonatal arterial switch operation and the unacceptable morbidity associated with atrial baffle operations, this operation has been accepted as the procedure of choice for the treatment of complete transposition of the great arteries. It restores the left ventricle to its natural systemic function. Long-term sur-vival exceeds 90%. Translocation of the coronary arteries remains one of the most difficult aspects of the operation and late mortality appears to coincide with coronary artery events with sudden death secondary to acute myocardial infarction being reported in 1-2% of hospital survivors. Supravalvar pulmonary stenosis, neoaortic root dilation and valvar regurgitation and myocardial perfusion abnor-malities are aspects which require routine follow up [10]. Death hazard rate is extremely low by 6 to 12 months after an arterial switch operation and survival declined minimally after that time.

In the third world countries, where timely referrals are difficult, there still is a role for the Sennings / Mustard operations. The cardiac problem to be followed in these cases will be the failure of systemic right ventricle and systemic tricuspid valve regurgitation (pronounced more if repair was for TGA + VSD). Atrial arrhythmias are the next most long term issue after these types of surgeries. A review of 40 years of experience of the atrial switch results highlighted 75% 25 year survival. Late morbidity included issues mentioned above. Progressive heart failure necessitating surgery or late sudden death from arrhythmias are the main etiologies of late death [11].

TOTAL ANOMALOUS PULMONARY VENOUS RETURN (TAPVR)

Obstructed variety of TAPVR is an absolute surgical emergency and requires immediate correction. Other forms will require a surgical correction by 3-6 months of life or whenever diagnosed.

With timely correction this lesion has an excellent long term outcome. During follow up pulmonary venous obstruction, supraventricular arrhythmias and sinus node dysfunction should be sought for Associated heterotaxy carries very poor prognosis.

INTERVENTIONAL PROCEDURES

Balloon valvuloplasty

Valvuloplasty is usually performed for aortic and the pulmonary valves. If the diagnosis is made in the newborn period and associated ventricular dysfunction, valvulo-plasty needs to be performed in all these patients. Indications that the obstruction is critical is that the cardiac output is compromised or the cardiac function is affected or if the gradient is very high and the valve opening is limited to a pin hole [12]. Recent publications support pulmonary valvuloplasty for gradients of 50 mmHg or more for best outcomes. These patients carry an excellent long term prognosis uniformly. Newborn pulmonary stenosis carries a higher recurrence rate of 10%.

Aortic valvuloplasty becomes emergent when in neonatal period the ventricular function is depressed due to

the obstruction. In non-neonatal period, a gradient above 60 mmHg is taken as an indicator for intervention unless more than trivial aortic regurgitation is present. Long term outlook is not as good as pulmonary valve intervention with 50% patients who are intervention free over an 8 year period [13].

DEVICE CLOSURE

Device closure is a preferred modality for closure of patent ductus arteriosus, atrial septal defect and muscular ventricular septal defect. For perimembranous ventricular septal defects, there have been several reports but the complications continue to be high and procedure technically challenging.

Patent ductus arteriosus closure with devices can be performed in children 5 Kg and more with a significant level of ease and comfort without any longterm effects. This has been substantiated in recent publications.

Atrial Septal Defect closure has been accomplished with the Amplatzer devices for appropriately selected ASD's without complications. Only those ASD's with the Aortic rim missing or very large ASD's with small rims have a higher embolization of device or late aortic perforation.

Muscular ventricular septal defects with significant shunting and moderate pulmonary hypertension require closure when diagnosed if the child is beyond 8-10 Kg. Most often, large muscular VSD's require surgery before they reach 8-10 kgs. Few patients truly fit the criteria for device closure of muscular VSD's. Even multiple muscular VSD's can be closed with a single device.

EMERGENCY INTERVENTIONS

Most often indicated emergency intervention is balloon atrial septostomy. This is usually performed for transposition of great arteries. This is performed in early neonatal period prior to preparation of the arterial switch operation.

In addition several interventions may have to be performed as an emergency in post operative patients. This is rare though in contemporary practice but a rare BT shunt thrombosis, early re-coarctation may have to be intervened on. Fontan with fenestration may require re-opening of a closed fenestration in the early post-op period. Similarly, Glenn operations, a venous shunt, may also thrombose and require catheter directed thrombolytic therapy.

Finally, embolized devices or central lines may need to be removed as an emergency. Bleeding collaterals may need to be closed emergently with coils to stop hemoptysis.

Tamponade usually needs to be drained emergently and this is also performed in the cath lab or at the bedside.

REFERENCES

1. Lammers A, Hager A, Eicken A, Lange R, Hauser M, Hess J. Need for closure of secundum atrial septal defect in infancy. J Thorac Cardiovasc Surg 2005; 129(6): 1353-1357.
2. Jost CH, Connolly HM, Danielson GK, Bailey KR, Schaff HV, Shen WK, Warnes CA, Seward JB, Puga FJ, Tajik AJ. Sinus venosus atrial septal defect: long-term postoperative outcome for 115 patients. Circulation 2005;112(13):1953-1958.
3. Gersony WM. Long-term follow-up of operated congenital heart disease. Cardiol Clin 1989; 7(4): 915-923.
4. Bando K, Turrentine MW, Sun K, Sharp TG, Ensing GJ, Miller AP, Kesler KA, Binford RS, Carlos GN, Hurwitz RA, *et al*. Surgical management of complete atrioventricular septal defects. A twenty-year experience. J Thorac Cardiovasc Surg. 1995; 110(5): 1543-1552.
5. Toro-Salazar OH, Steinberger J, Thomas W, Rocchini AP, Carpenter B, Moller JH. Long-term follow-up of patients after coarctation of the aorta repair. Am J Cardiol 2002; 89(5): 541-547.
6. Ullmann MV, Gorenflo M, Sebening C, Ulmer HE, Hagl S. Long-term results after repair of truncus arteriosus communis in neonates and infants. Thorac Cardiovasc Surg 2003; 51(4): 175-179.
7. Gatzoulis MA, Elliott JT, Guru V, Siu SC, Warsi MA, Webb GD, Williams WG, Liu P, McLaughlin PR. Right and left ventricular systolic function late after repair of tetralogy of Fallot. Am J Cardiol 2000; 86(12): 1352-1357.
8. Gatzoulis MA, Balaji S, Webber SA, Siu SC, Hokanson JS, Poile C, Rosenthal M, Nakazawa M, Moller JH, Gillette PC, Webb GD, Redington AN. Risk factors for arrhythmia and sudden cardiac death late after repair of Tetralogy of Fallot: a multicentre study. Lancet 2000; 356(9234): 975-981.
9. Earing MG, Cetta F, Driscoll DJ, Mair DD, Hodge DO, Dearani JA, Puga FJ, Danielson GK, O'Leary PW. Long-term results of the Fontan operation for double-inlet left ventricle. Am J Cardiol 2005; 96(2): 291-298.
10. Massin MM. Midterm results of the neonatal arterial switch operation. A review. J Cardiovasc Surg (Torino) 1999; 40(4): 517-522.
11. Oechslin E, Jenni R. Forty years after the first atrial switch procedure in patients with transposition of the great arteries: long-term results in Toronto and Zurich. Thorac Cardiovasc Surg 2000 Aug; 48(4): 233-237.
12. Latson LA. Critical pulmonary stenosis. J Interv Cardiol 2001 Jun; 14(3): 345-350.
13. Latiff HA, Sholler GF, Cooper S. Balloon dilatation of aortic stenosis in infants younger than 6 months of age: Intermediate outcome. Pediatr Cardiol 2003 Jan-Feb; 24(1):17-26.