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Congenital Heart Defects – A Review

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1. Introduction

Congenital heart defect (CHD) may be defined as an anatomic malformation of the heart or great vessels which occurs during intrauterine development, irrespective of the age at presentation. Ventricular septal defect and coarctation of the aorta are typical examples of CHDs. In this chapter, a brief review of incidence, etiology and classification of CHD, and an overview of the most common congenital cardiac anomalies and their management will be presented. Cardiac abnormalities, generally considered not congenital in origin but important cardiac problems in children, namely rheumatic heart disease, Kawasaki syndrome and cardiomyopathy will not be discussed in this review. Also, discussion of important symptoms/findings/issues with which the children are referred to pediatric cardiologists such as cardiac murmur, chest pain, syncope/dizziness, palpitation, arrhythmia, hypertension, clearance for participation in sports, coronary risk factors, bacterial endocarditis prophylaxis, ADHD medication use, clearance for non-cardiac surgery and others will not be included in the this chapter.

Incidence of congenital heart defects

The reported incidence of congenital cardiac defects varies between 0.47 to 1.17% of live births, but 0.6% to 0.8% of live births is considered typical. This would result in birth of 25,000 to 35,000 infants with CHD each year in the United States alone. Congenital heart defects are more common than well-known congenital anomalies such as congenital pyloric stenosis, cleft lip, Down syndrome and congenital dislocation of the hip.

2. Etiology

The exact cause of all congenital cardiac defects is not known. The majority of the defects can be explained by multifactorial inheritance hypothesis (Nora 1968) which states that a predisposed fetus, when exposed to a given environmental trigger (to which the fetus is sensitive) during the critical period of cardiac morphogenesis will develop the disease. This genetic and environmental interaction is most likely to be pathogenetic mechanism of congenital heart defects. Calculations based on this hypothesis predict the frequency of occurrence of the disease in first degree relatives to be square root of its frequency in the population; this fits the congenital heart disease figures (Nora 1968).

A variety of factors have statistical association with certain heart defects and these may be termed risk factors. Maternal rubella appears to have causative association with heart defects. Significantly higher incidence of serologic evidence for Coxsackie B virus infection

during pregnancy in mothers of infants with congenital heart defects than in matched-control women suggested causative relationship between Coxsackie B infection and congenital heart defects, but this evidence is neither conclusive nor confirmed. Among drugs, maternal ingestion of thalidomide during pregnancy is associated with high incidence heart defects in the offspring. Similar association has been reported for some anticonvulsant drugs (particularly dilantin and trimethadione), alcohol (excessive), Lithium, sex hormones, diazepam, carticosteroids, phenolhiazine, folic acid antagonists, cocaine and dextromethamphetamines. A higher incidence of cardiac abnormalities with maternal diabetes is well known. Gross chromosomal anomalies such as trisomy 21 (Down syndrome), trisomy D and E syndromes, Turner's syndrome (XO), partial deletion of chromosome 22 and cri-du-chat (partial dilation of the short arm of chromosome 5) are associated with a higher incidence of heart defects than normal population and are likely be responsible for the congenital heart defects. Some generalized syndromes, secondary to a single mutant gene (for example, Marfan) involving multiple organ systems are associated with cardiovascular defects peculiar to that particular syndrome (Rao 1977a). Both autosomal (dominant and recessive) and sex-linked (dominant and recessive) single mutant gene syndromes have been reported with CHD. Finally, less than 1% of congenital heart defects can be explained by simple Mendelian inheritance. Autosomal dominant transmission other than single mutant gene syndrome has been reported with atrial septal defect, patent ductus arteriosus, aortic stenosis, pulmonary stenosis, tetralogy of Fallot and hypertrophic cardiomyopathy. Autosomal recessive inheritance may be present in some forms of endocardial fibroelastosis. To the best of my knowledge, sex-linked transmission has not been reported with CHD. However, recently questions have been raised as to the mitochondrial inheritance in which maternal transmission to almost all offspring occurs. In the presence of family history of congenital heart defect (parent or sibling) the probability of CHD in the offspring is higher than that seen in general population.

In summary, the cause of congenital heart defects is largely unknown and the majority of them may be explained by multifactorial inheritance hypothesis. Extensive research on gene mapping that is currently in progress may unravel previously unknown genetic mechanisms for CHD. Also, several chapters to follow address the issues related to causation of CHD.

3. Classification

Congenital heart defects may be classified into acyanotic and cyanotic depending upon whether the patients clinically exhibit cyanosis. The acyanotic defects may further be subdivided into obstructive lesions and left-to-right shunt lesions. The cyanotic defects, by definition, have right-to-left shunt. The relative incidence of these groups of defects and most common defects in each group are listed in table I. The total percent is not 100 because some of the heart defects cannot be classified into the categories listed.

4. Ayanotic heart defects: Obstructive lesions

When there is a significant narrowing of a valve or a blood vessel, there is a higher pressure proximal to the obstruction compared to the distal pressure; this pressure gradient is necessary to maintain flow across the stenotic site. Hypertrophy of the cardiac chamber proximal to the obstruction and flow disturbance across the site of obstruction and their effects will determine the clinical features.

Type of Defect	% incidence
Acyanotic	65%
Obstructive Lesions Pulmonary stenosis Aortic stenosis Coarctation of the aorta	25%
Left-to-right shunt lesions Atrial septal defect Ventricular septal defect Patent ductus arteriosus	40%
Cyanotic Tetralogy of Fallot Transposition of the great arteries Tricuspid atresia	20%

Table 1. Classification of Congenital Heart Defects

4.1 Pulmonary stenosis

The obstruction can be at valvar, subvalvar or supravalvar sites or in the branch pulmonary arteries (Rao 2000a). Valvar stenosis is the most common type and will be discussed in this section. Valvar pulmonary stenosis (PS) constitutes 7.5% to 9.0% of all CHDs. The pathologic features of valvar stenosis vary, but the most commonly found pathology is what is described as "dome shaped" pulmonary valve with fusion of the thickened pulmonary valve leaflets. Hypertrophy of the right ventricle (proportional to the degree of obstruction) and dilatation of main pulmonary artery (not related to the severity of obstruction) are also seen.

4.1.1 Symptoms

Children with PS usually present with asymptomatic murmurs, although they can present with signs of systemic venous congestion (usually interpreted as congestive heart failure) due to severe right ventricular dysfunction or cyanosis because of right-to-left shunt across the atrial septum.

4.1.2 Physical findings

The right ventricular and the right ventricular outflow tract impulses are increased and a heave may be felt at the left lower and upper sternal borders. A thrill may be felt at the left upper sternal border and/or in the suprasternal notch. The first heart sound may be normal or loud. The second heart sound is variable, depending upon the degree of obstruction and will be detailed later in this section. An ejection systolic click is heard in most cases of valvar stenosis. The click is heard best at the left lower, mid and upper sternal borders and varies with respiration (decreases or disappears with inspiration). An ejection systolic murmur (Figure 1, top) is heard best at the left upper sternal border and it radiates into infraclavicular regions, axillae and back. The intensity of the murmur may vary between grades II-V/VI; the intensity is not necessarily related to the severity of the stenosis.

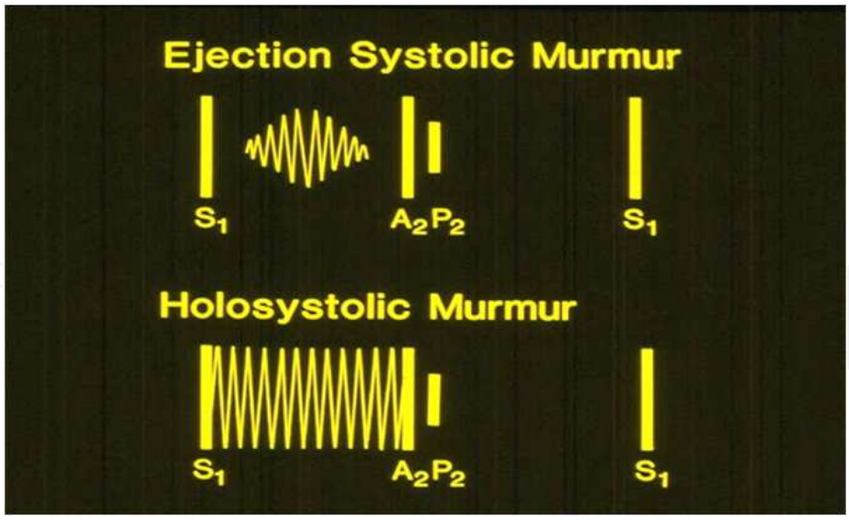

Fig. 1. Auscultatory diagrams of systolic murmurs. Ejection systolic murmur (top) begins shortly after the first heart sound (S_1) and ends shortly before the second heart sound (A_2, aortic component and P_2, pulmonary component) whereas a holosystolic murmur (bottom) begins with and obscures the S_1 and may last throughout the systole (as in the diagram) or may stop short of A_2.

4.1.3 Clinical assessment of severity

The timing of the click, the extent of splitting of the second sound, the intensity of the pulmonary component of the second sound, the length (duration) of the murmur, and timing of peaking of the systolic murmur are usually suggestive of the severity of pulmonary valve obstruction (Figure 2) (Rao 1991b, Vogelpoel & Schriere 1960).

The loudness of the ejection systolic murmur does not indicate the severity of obstruction but rather its duration and time of peaking; the longer the murmur and the later it peaks, the more severe is the PS. Similarly, the shorter the time interval between the first heart sound and ejection click, the wider the splitting of the second heart sound, and softer the pulmonary component, the more severe is the degree of pulmonary valve obstruction (Rao 2000).

4.1.4 Noninvasive evaluation

4.1.4.1 Chest x-ray

In most cases, the chest film shows no cardiomegaly, but a characteristically dilated main pulmonary artery segment (post-stenotic dilatation) is visualized. The magnitude of pulmonary artery dilatation has no bearing on the severity of pulmonary valve stenosis.

4.1.4.2 Electrocardiogram (ECG)

The ECG shows right ventricular hypertrophy; the degree of right ventricular hypertrophy is proportional to the severity of stenosis. Right atrial enlargement may be present.

4.1.4.3 Echocardiogram

The echo may show right ventricular enlargement without paradoxical septal motion and thickened and domed pulmonary valve leaflets. The Doppler flow velocity across the site of obstruction is increased and the magnitude of this increase reflects the severity of

pulmonary valve stenosis. The peak instantaneous pressure gradient can be calculated by the use of a modified Bernoulli equation:

$$\Delta P = 4 V^2$$

Where, Δ P is peak instantaneous pressure gradient in mmHg and V is the peak velocity across the valve in meters/sec.

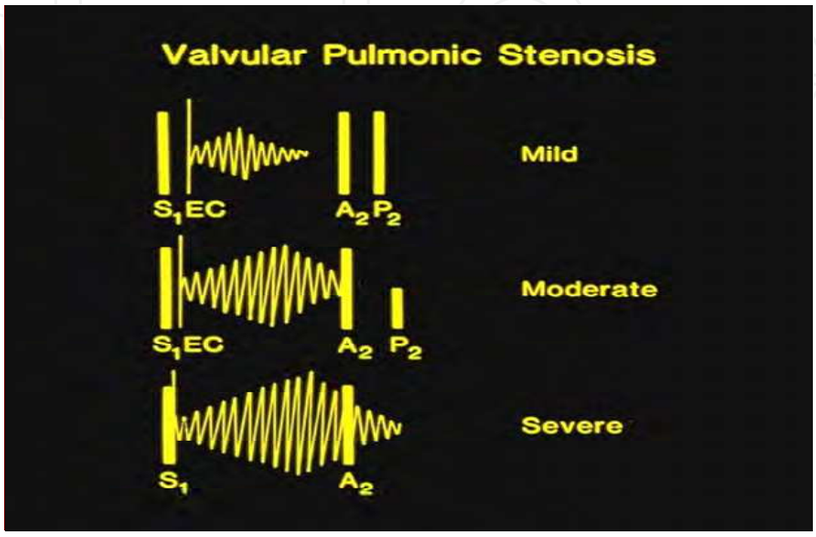

Fig. 2. In valvar pulmonary stenosis, severity of obstruction may be judged by auscultatory findings. In mild cases of pulmonary valve stenosis, the click (EC) is clearly separated from the first heart sound, almost normal splitting of the second heart sound with normal or slightly increased pulmonary component (P_2) of the second sound is heard, and an ejection systolic, diamond-shaped murmur that peaks early in systole and ends way before the aortic closure of the second heart sound is appreciated. The findings in moderate PS include an ejection systolic click that is much closer to the first heart sound than in milder forms, widely split second sound with diminished pulmonary component of the second sound and an ejection systolic murmur that peaks in mid to late systole and ends just below the aortic component (A_2) of the second sound. The features of severe valvar PS are an ejection systolic click which is either not present or falls so close to the first heart sound that it becomes inseparable from it, markedly increased splitting with a soft or inaudible pulmonary component of the second heart sound, and a long ejection systolic murmur that peaks late in systole and extends beyond the aortic component of the second sound so that the latter cannot be heard.

4.1.5 Cardiac catheterization and selective cineangiography

Though this procedure is not required for diagnosing valvar PS, it is usually required prior to therapeutic intervention, to be discussed below. The oxygen saturation data usually do not show evidence for left-to-right shunts. A right-to-left shunt across the patent foramen ovale (or an atrial defect) may be present in moderate to severe pulmonary valve obstruction. Right atrial pressure (particularly 'a' wave) may be increased. The right ventricular peak systolic pressure is increased. Trans-pulmonary valve peak-to-peak gradient is indicative of severity of obstruction. A peak-to-peak gradient in excess of 50 mmHg is usually considered an indication for therapeutic intervention. Angiocardiography

usually reveals thickened and domed pulmonary valve leaflets with a thin jet of passage of contrast across the pulmonary valve. Enlargement of the right ventricle and dilated main pulmonary artery segment are also seen. In patients with severe or long-standing pulmonary valve obstruction, infundibular constriction may be seen.

4.1.6 Natural history

The natural history studies (Nugent et al 1977) have classified the degree of pulmonary valve obstruction based on peak-to-peak catheter-measured pulmonary valvar gradient: trivial = gradient < 25 mmHg; mild = gradient 25-49 mmHg; moderate = gradient 50 to 79 mmHg and severe = gradient > 80 mmHg. Patients with trivial and mild (gradients < 50 mmHg) pulmonary stenosis generally remain mild at follow-up. Patients with moderate stenosis (gradients of 50 to 79 mmHg) in contradistinction to trivial and mild stenosis, progressively increase their gradient

4.1.7 Management

Until early 1980s, surgical pulmonary valvotomy was the only treatment available, but at the present time relief of pulmonary valve obstruction can be accomplished by balloon pulmonary valvuloplasty. Indeed, at the present time balloon pulmonary valvuloplasty is treatment of choice. The indications for intervention are similar to those prescribed for surgery: a peak-to-peak systolic pressure gradient > 50 mmHg across the pulmonary valve with a normal cardiac index (Rao 1988, Rao 1989b, Rao 1998). Detailed description of the procedure of balloon valvuloplasty and the results of such a procedure are beyond the scope of this chapter; the reader is referred elsewhere for these details (Rao 2007a, Rao 2007b). In brief, a balloon catheter (with a deflated balloon) is positioned across the pulmonary valve and the balloon inflated (Figure 3); the radial forces of balloon inflation produce valve leaflet commissural disruption and thus relief of pulmonary valve obstruction (Rao 1993).

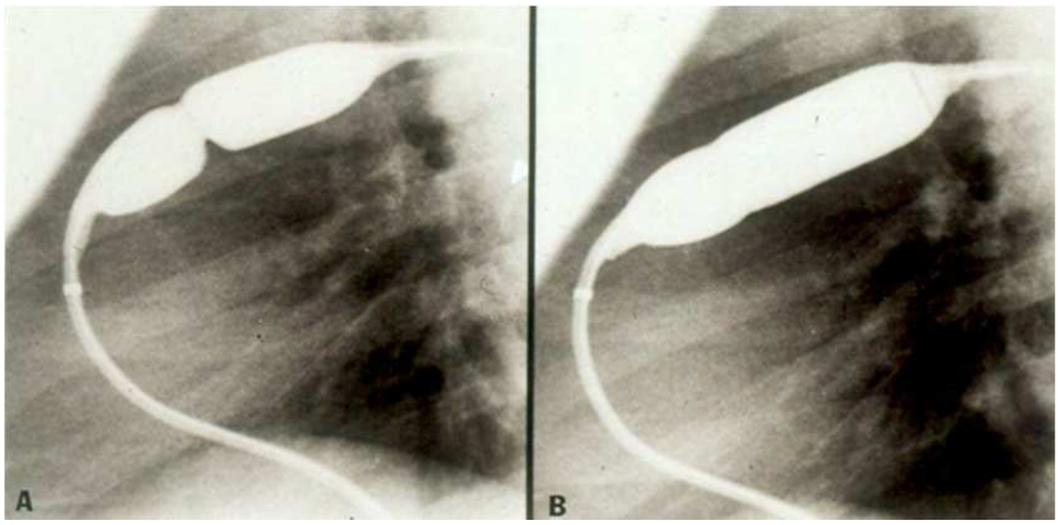

Fig. 3. Selected cineradiographic frames of a balloon dilatation catheter placed across the pulmonary valve. Note "waisting" of the balloon during the initial phases of balloon inflation (A), which is almost completely abolished during the later phases of balloon inflation (B). Reproduced with permission of the author and publisher, Rao PS: Transcatheter Therapy in Pediatric Cardiology, Wiley-Liss, Inc, New York, 1993, p. 62.

Previous recommendations are to use a balloon that is 1.2 to 1.4 times the size of the pulmonary valve annulus; however, more recent recommendations are to limit the balloon/annulus ratio to 1.2 to 1.25 (Rao 2000b, Rao 2007a, Rao 2007b). When the pulmonary valve annulus is too large to dilate with a single balloon, valvuloplasty with simultaneous inflation of two balloons across the pulmonary valve annulus is recommended. Immediate, short-term and long-term results (Figure 4) are good; although long-term results are limited (Rao et al 1998).

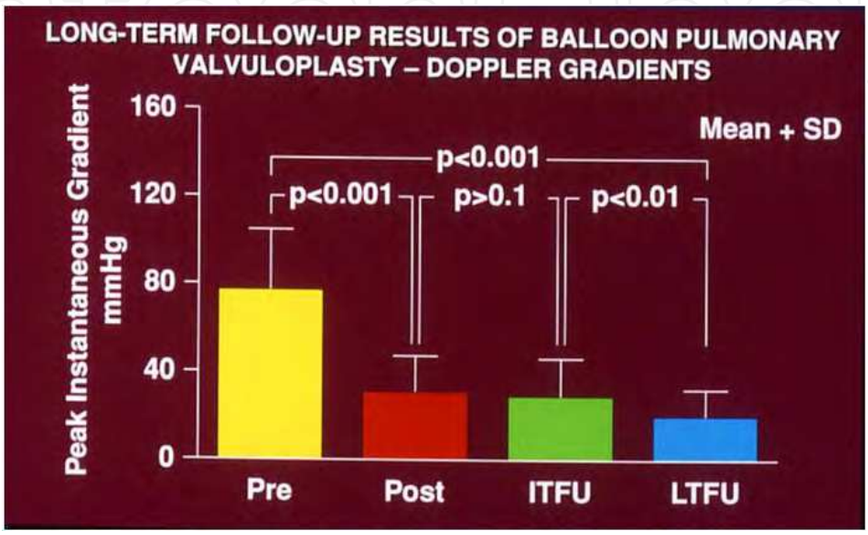

Fig. 4. Bar graph showing maximum peak instantaneous Doppler gradients, indicative of severity of pulmonary stenosis, prior to (Pre), one day following (Post) balloon pulmonary valvuloplasty and at intermediate-term (ITFU) and late (LTFU) follow-up. Note significant reduction ($p < 0.001$) after valvuloplasty, which remains unchanged ($p > 0.1$) at ITFU. However, at LTFU there was further fall ($p < 0.01$) in the Doppler gradients.

Given the success with balloon pulmonary valvuloplasty, surgery is reserved for unsuccessful balloon cases, mostly for cases with supravalvar PS, severe valve annular hypoplasia and dysplastic pulmonary valves.

In patients with mild pulmonary valve stenosis, periodic clinical follow-up, antibiotic prophylaxis prior to any bacteremia-producing procedures to prevent subacute bacterial endocarditis and no exercise restriction are recommended.

4.2 Aortic stenosis

Left ventricular outflow tract obstruction may occur at valvar, subvalvar (fixed subaortic stenosis and idiopathic hypertrophic subaortic stenosis) and supravalvar locations (Singh and Rao 2009). Valvar stenosis is the most common form and will be discussed in this section. The prevalence of congenital valvar aortic stenosis (AS) is 5% to 6% of patients with CHD. The pathology of the stenotic aortic valve is variable, most commonly it is a bicuspid valve with varying degrees of commissural fusion of thickened, domed, nonpliable valve leaflets. Tricuspid and rarely unicuspid aortic valve leaflets can also cause aortic valve obstruction. Dysplasia of the aortic valve leaflets with or without hypoplasia of the valve ring may be found in neonates and young infants. Calcification of

the aortic valve leaflets so frequent in the elderly is uncommon during childhood. Dilatation of ascending aorta, post-stenotic dilatation, is seen in most cases, and the extent of aortic dilatation is independent of the severity of aortic obstruction. Hypertrophy of the left ventricular muscle is concentric in nature and is largely proportional to the degree of obstruction.

4.2.1 Symptoms

The majority of children with valvar AS are asymptomatic and the AS is detected because of a cardiac murmur heard on routine auscultation. When symptoms are exhibited, dyspnea, easy fatigability or chest pain is presenting complaint. Syncope may be a presenting complaint in some children with severe AS. In contradistinction to children, neonates and young infants usually present with dyspnea and signs of heart failure.

4.2.2 Physical findings

The left ventricular impulse is increased (left ventricular heave) in all but mild cases. A thrill may be felt at the right upper sternal border and/or in the supra-sternal notch. The first heart sound is usually normal. The second heart sound is also normal unless the aortic stenosis is extremely severe when there may be a paradoxical splitting of the second heart sound. An ejection systolic click is heard best at the apex and left mid and right upper sternal borders and the click does not vary with respiration. An ejection systolic murmur of grade II-V/VI intensity is usually heard best at the right upper sternal border with radiation into both carotid arteries. The arterial pulses are usually normal.

4.2.3 Noninvasive evaluation

4.2.3.1 Chest roentgenogram

In most cases, the chest X-ray shows a normal sized heart and a dilated ascending aorta; the latter is a sign of post-stenotic dilatation. In neonates and those with very severe heart failure cardiomegaly is seen.

4.2.3.2 Electrocardiogram

The ECG may be normal or may show varying degrees of left ventricular hypertrophy. Inverted T waves in the left chest leads indicate that aortic valve obstruction is severe. However, not all severe AS patients show T wave inversion.
None of the above described clinical and laboratory data have any predictive value in determining the severity of aortic valve obstruction.

4.2.3.3 Echocardiogram

The echocardiogram may show thickened and domed aortic valve leaflets. The aortic valve is usually bicuspid (Figure 5), with eccentric opening.
The left ventricular muscle may be thickened and its shortening fraction may be increased, depending upon the severity of AS. Doppler flow velocity across the aortic valve is increased and can be used to quantitate peak instantaneous gradient across the aortic valve in a manner similar to that described for the pulmonary valve. However, Doppler-derived mean systolic gradient appears to reflect peak-to-peak catheter gradient (see below) more accurately than peak instantaneous Doppler gradient. Mild degree of aortic insufficiency

may be seen by color Doppler, even in patients without auscultatory evidence for aortic regurgitation.

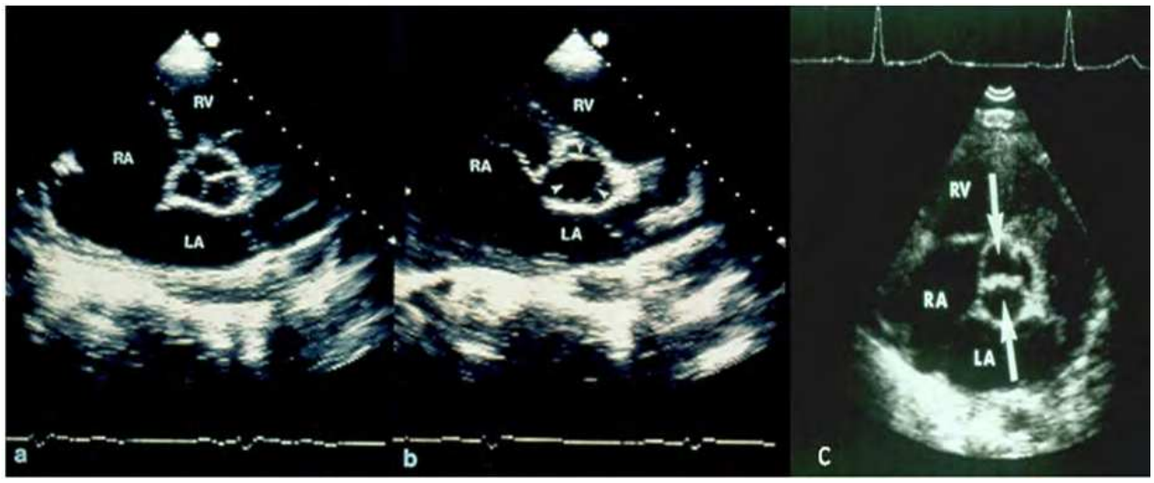

Fig. 5. Short axis views of the aorta showing aortic valve leaflets in closed (a) and open position (b) in children with tricuspid aortic valves (a and b). Bicuspid aortic valve (large arrows) is shown in c, which is commonly associated with aortic stenosis. Three aortic valve cusps and commissures (in a) are clearly seen and contrast with two valve cusps and single horizontal commissure (in c). Arrow heads in b point to open aortic valve leaflets. Neither of the children showed clinical or echo-Doppler evidence for aortic stenosis and are shown here only to demonstrate the bicuspid and tricuspid valve leaflets. LA, left atrium; RA, right atrium; RV, right ventricle.

4.2.4 Cardiac catheterization and angiography

The data show elevated left ventricular peak systolic pressure with a peak-to-peak pressure gradient across the aortic valve indicative of the severity of obstruction. Angiography will confirm thickened domed aortic valve leaflets and exclude any other abnormalities.

4.2.5 Management

The indications for intervention in valvar AS is a peak-to-peak gradient >50 mmHg with either symptoms or electrocardiographic ST-T wave changes or a peak gradient >70 mmHg irrespective of symptoms or ECG changes (Rao 1989b, Rao 1990). When pressure gradients are used as criteria for intervention (instead of valve area), it must be assured that the cardiac index is normal during pressure measurement. Until recently, surgical commissurotomy was the treatment of choice. Since the introduction of balloon valvuloplasty for valvar AS in 1983, increasing number of pediatric cardiologists, including the author of this chapter have been using balloon aortic valvuloplasty as a first therapeutic procedure for relief of aortic valve obstruction although, at this time, there is no consensus with regard to the choice of treatment mode. When surgical commissurotomy is chosen it is usually performed on cardiopulmonary bypass. When balloon valvuloplasty is performed, a

balloon diameter size 80% to 100% of the size of the aortic valve annulus is chosen for valvuloplasty (Rao 1990). Immediate, short-term and long-term results following balloon aortic valvuloplasty (Figure 6) are encouraging. Only limited long-term results are available to-date (Galal et al 1997, Rao 1999).

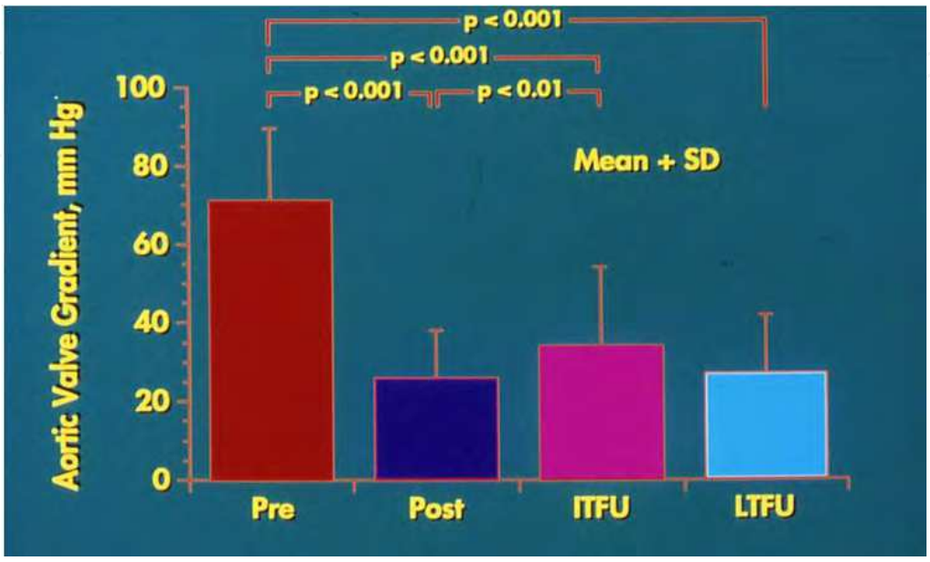

Fig. 6. Bar graph showing maximum peak instantaneous Doppler gradients, indicative of severity of aortic stenosis, prior to (Pre), one day following (Post) balloon aortic valvuloplasty and at intermediate-term (ITFU) and late (LTFU) follow-up. Note significant reduction ($p < 0.001$) after valvuloplasty, which continues to be lower ($p < 0.001$) at ITFU and LTFU.

For milder forms of AS, subacute bacterial endocarditis prophylaxis and periodic follow-up are necessary. Restriction from participation in competitive sports is recommended for all but mildest form of AS.

4.3 Coarctation of the aorta

The prevalence of coarctation of the aorta (CoA) was found to vary between 5% and 8% of CHDs; however, coarctation may be found more frequently in infants presenting with symptoms prior to one year of age. In the past, CoA was designated as preductal (or infantile) or postductal (or adult) type, depending on whether the coarctation segment was proximal or distal to the ductus arteriosus, respectively. However, a closer examination of the anatomy suggests that all coarctations are juxtaductal. The coarctation may be discrete, or a long segment of the aorta may be narrowed; the former is more common. Classic CoA is located in the thoracic aorta distal to the origin of the left subclavian artery, at about the level of the ductal structure. However, rarely, a coarcted segment may be present in the abdominal aorta. Varying degrees of hypoplasia of the isthmus of the aorta (the portion of the aorta between the origin of the left subclavian artery and the ductus arteriosus) and transverse aortic arch (the arch between the origin of the innominate artery and the left

subclavian artery) are present in the majority of patients with CoA; this hypoplasia may be significant in symptomatic CoA of the neonate and infant, whereas in older children there may be only a mild degree of narrowing. The most commonly associated defects are patent ductus arteriosus, ventricular septal defect and AS. The younger the infant presents, the more likely that there is a significant associated defect. Bicuspid aortic valve and abnormal mitral valve are also seen. Sometimes, CoA is a complicating feature of a more complex, cyanotic heart defects, such as transposition of the great arteries, Taussig-Bing anomaly, double-inlet left ventricle, tricuspid atresia with transposition of the great arteries, and hypoplastic left heart syndrome. In this section, I will discuss CoA in children older than 1 year of age.

4.3.1 Symptoms

Children beyond infancy usually are asymptomatic; an occasional child will complain of pain or weakness in the legs. Most often, the coarctation is detected because of a murmur or hypertension which is detected on a routine examination (Rao 1995).

4.3.2 Physical findings

A clinical diagnosis of CoA is best made by simultaneous palpation of femoral and brachial pulses. The left ventricular impulse may be increased. A thrill is usually felt in the supra-sternal notch. The first and second heart sounds are usually normal in isolated aortic coarctation. Since a large percentage (up to 60%) of patients with CoA have associated bicuspid aortic valves, an ejection systolic click may be heard at right upper and left mid sternal borders and apex; this click does not change with respiration. An ejection systolic murmur may be heard at left or right upper sternal borders, but is usually heard best over the back in the inter-scapular regions. Sometimes a continuous murmur may be heard in the left inter-scapular region secondary to continuous flow in the coarcted segment or on the back (secondary to flow in the collateral vessels). Palpation of the brachial and femoral artery pulses simultaneously will reveal delayed and decreased or absent femoral pulses. Blood pressure in both arms and one leg must be determined: a peak systolic pressure difference of more than 20 mmHg in favor of arms may be considered as evidence for coarctation of the aorta (Rao 1995). Involvement of the left subclavian artery in the coarctation or anomalous origin of the right subclavian artery (below the level of coarctation) may produce decreased or absent left or right brachial pulses, respectively, and therefore palpation of both brachial pulses and measurement of blood pressure in both arms are important.

4.3.3 Noninvasive evaluation

4.3.3.1 Chest x-ray

Chest roentgenogram may show a normal sized heart or the heart may be mildly enlarged. Other roentgenographic features include a "3" sign on a highly penetrated chest x-ray, inverted "3" sign of the barium filled esophagus and rib-notching (secondary to collateral vessels).

4.3.3.2 Electrocardiogram

The ECG may be normal or may show left ventricular hypertrophy.

4.3.3.3 Echocardiogram

Echocardiographic studies usually reveal the coarctation in the supra-sternal notch, two-dimensional echo views of the aortic arch. Increased Doppler flow velocity in the descending aorta by continuous-wave Doppler and demonstrable jump in velocity at the coarcted segment by pulsed-Doppler technique are usually present. Extension of the Doppler flow signal into the diastole is indicative of significant obstruction. Instantaneous peak pressure gradients across the aortic coarctation can be calculated by employing modified Bernoulli equation in manner similar to that described for PS and AS. Because of higher proximal velocity, coarctation gradients may be more accurately estimated by:

$$\Delta P = 4 (V_2^2 - V_1^2)$$

Where, ΔP is peak instantaneous gradient and V_2 and V_1 are peak Doppler velocities in the descending aorta distal to the coarctation (continuous wave Doppler) and proximal to the coarctation (pulsed Doppler), respectively.
But the calculated gradient is usually an over-estimation, especially if there is no diastolic extension of the Doppler velocity (Rao and Carey 1989).

4.3.4 Catheterization and angiography

In isolated aortic coarctation, elevation of left ventricular and ascending aortic peak systolic pressure with significant peak-to-peak systolic pressure gradient across the coarctation is found. Selective aortic root or aortic arch angiography is necessary to clearly demonstrate the aortic narrowing.

4.3.5 Management

Significant hypertension and/or congestive heart failure are indications for intervention. In the presence of congestive heart failure, conventional anti-congestive measures including digitalis and diuretics should be promptly instituted. In the presence of hypertension, it is better to relieve the obstruction promptly rather than attempting to "treat" hypertension with antihypertensive drugs. Aortic coarctation may be relieved either by surgery or by balloon angioplasty. Symptomatic children should undergo relief of coarctation soon after the child is stabilized. Asymptomatic children should undergo the procedure electively. If neither hypertension nor heart failure is present, elective relief of the obstruction between the ages of 2 and 5 years is suggested. Waiting beyond 5 years is not advisable because of evidence for residual hypertension if the aortic obstruction is not relieved by the age of 5 years.
Surgical relief of aortic coarctation is the conventional treatment option. Since the description balloon angioplasty in 1983, increasing number of cardiologists, including our group, have used this technique for relief of aortic coarctation (Rao 1989c; Rao et al 1996). While I believe that balloon angioplasty is the treatment option of choice for relief of native aortic coarctation, because of concern for development of aneurysms, some cardiologists prefer surgery. Balloon angioplasty may be an effective alternative to surgery for the relief of aortic coarctation. Children older than 1 year and adults with discrete native coarctation are candidates for balloon dilatation. Long-segment coarctations or those associated with significant isthmic hypoplasia may be candidates for stent placement, especially in adolescents and adults (Figure 7).

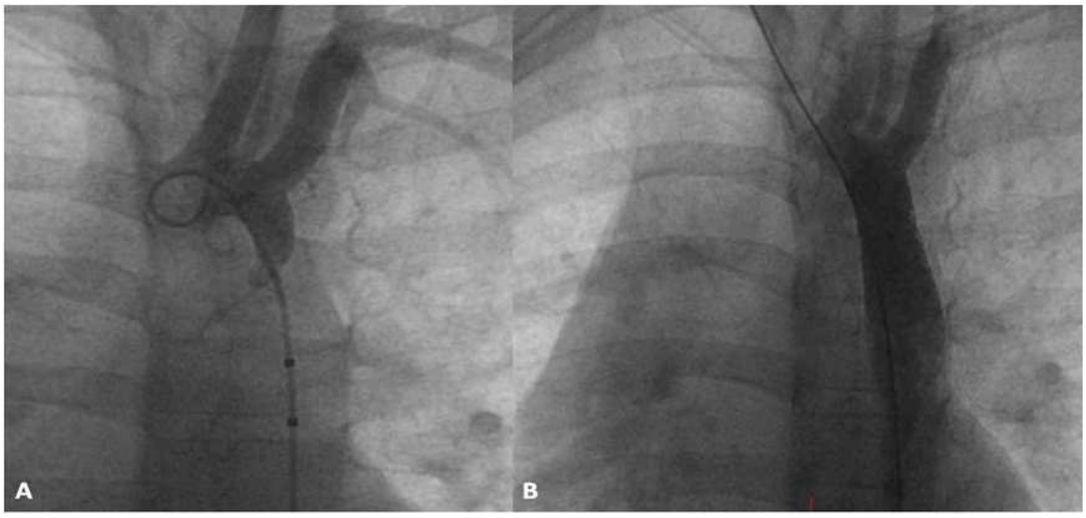

Fig. 7. Selected cine frames from aortic arch angiogram in 20-degree left anterior oblique projection demonstrating aortic coarctation with isthmic hypoplasia in an adolescent prior to (A) and immediately following (B) stent implantation.

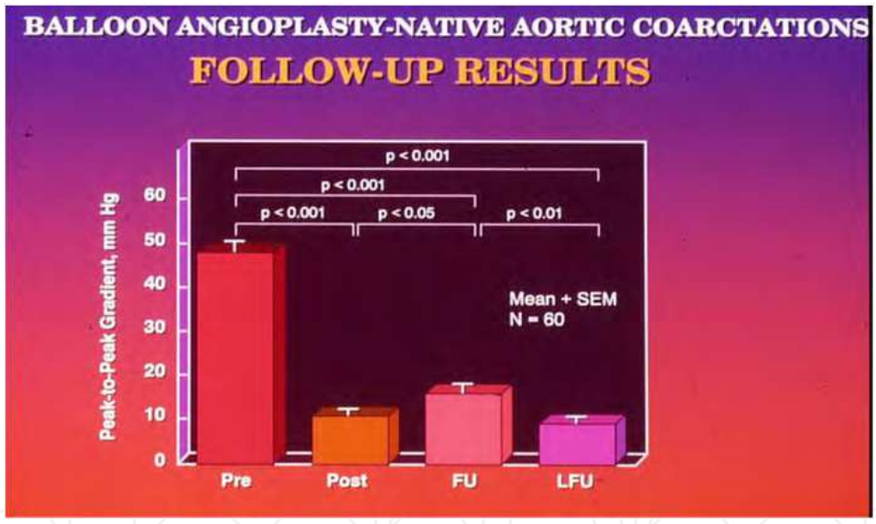

Fig. 8. Bar graph showing immediate and follow-up results after balloon angioplasty of native aortic coarctation. Peak-to-peak systolic pressure gradients across the coarctation in mmHg (mean + SEM) are shown. Note significant ($p < 0.001$) drop in the gradient following angioplasty (Pre, prior to vs. Post). The gradient increases ($p < 0.05$) slightly at a mean follow-up of 14 mo. However, these values are lower ($p < 0.001$) than those prior to angioplasty. At late follow-up (LFU), (median 5 years) following balloon angioplasty, blood pressure-measured arm-leg peak pressure difference is lower than catheterization measured peak gradients prior to ($p < 0.001$) balloon angioplasty and those obtained at intermediate-term follow-up ($p < 0.01$).

When surgical option is chosen, resection and end-to-end anastomosis, subclavian flap angioplasty or prosthetic patch angioplasty may be used depending upon anatomy of the aortic arch and coarctation and surgeon's preference. When balloon angioplasty is contemplated, the balloon size should be carefully chosen: the diameter of the balloon

should be two or more times the size of the coarcted segment, but no larger than the diameter of the descending aorta at the level of diaphragm. The immediate (Figures 8) and intermediate-term results of balloon coarctation angioplasty have been good although long-term follow-up is limited (Rao 1999).

5. Ayanotic heart defects: Left-to-right shunts

When there is a defect in the partition between left and right heart structures, the oxygenated blood is shunted from left-to-right because of generally lower pressure and/or resistance in the right heart than in the left. The physical findings are either a manifestation of flow across the defects or due to effects of excessive flow across the cardiac chambers (volume overload) and valves. The magnitude of the shunt determines the clinical presentation and symptoms.

5.1 Atrial septal defect

There are three major types of atrial septal defects (ASDs) and these include ostium secundum, ostium primum and sinus venosus defects. The clinical features are essentially similar but I will mainly concentrate on ostium secundum ASDs. Atrial septal defects constitute 8% to 13% of all CHDs. Pathologically, there is deficiency of the septal tissue in the region of fossa ovalis. These may be small to large. Most of the time, these are single defects, although, occasionally multiple defects and fenestrated defects can also be seen. Because of left-to-right shunting across the defects, the right atrium and right ventricle are dilated and somewhat hypertrophied. Similarly, main and branch pulmonary arteries are also dilated. Pulmonary vascular obstructive changes are not usually seen until adulthood.

5.1.1 Symptoms

Isolated ASD patients are usually asymptomatic and are usually detected at the time of preschool physical examination. Sometimes these defects are detected when echocardiographic studies are performed for some unrelated reason. A few patients do present with heart failure in infancy, although this is uncommon.

5.1.2 Physical examination

The right ventricular and right ventricular outflow tract impulses are increased and hyperdynamic. No thrills are usually felt. The second heart sound is widely split and fixed (splitting does not vary with respiration) and is the most characteristic sign of ASD. Ejection systolic clicks are rare with ASDs. The ejection systolic murmur of ASD is soft and is of grade I-II/VI intensity and rarely, if ever, louder. The murmur is secondary to increased flow across the pulmonary valve and is heard best at the left upper sternal border. A grade I-II/VI mid-diastolic flow rumble is heard (with the bell of the stethoscope) best at the left lower sternal border. This is due to large volume flow across the tricuspid valve. There is no audible murmur because of flow across the ASD.

5.1.3 Noninvasive evaluation

5.1.3.1 Chest x-ray

Chest film usually reveals mild to moderate cardiomegaly, prominent main pulmonary artery segment and increased pulmonary vascular markings.

5.1.3.2 Electrocardiogram

The ECG shows mild right ventricular hypertrophy; the so-called diastolic volume overload pattern with rSR' pattern in the right chest leads.

5.1.3.3 Echocardiogram

Echocardiographic studies reveal enlarged right ventricle with paradoxical septal motion, particularly well-demonstrable on M-mode echocardiograms. By two-dimensional echocardiogram, the defect can be clearly visualized (Figure 9A). The type of ASD, secundum versus primum can also be delineated by the echocardiographic study. Apical and precordial views may show "septal drop-outs" without an ASD because of thinness of the septum in the region of fossa ovalis. Therefore, only subcostal views should be scrutinized for evidence of ASD. In addition, demonstration of flow across the defect with pulsed Doppler (not shown) and color Doppler (Figure 9B) echocardiography is necessary to avid false positive studied. In adolescents and adults transesophageal echo is needed to make definitive diagnosis of ASD.

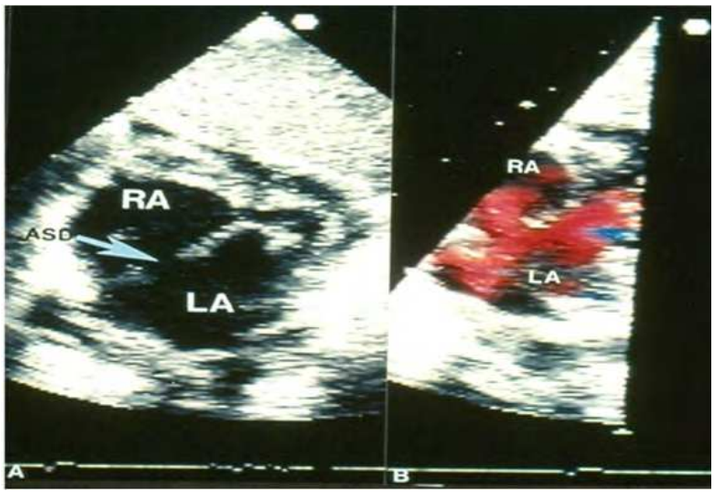

Fig. 9. Two dimensional subcostal echocardiographic view of the atrial septum (A) demonstrating a secundum atrial septal defect (ASD) in the mid septum (arrow). Color Doppler imaging shows left-to-right shunt. LA, left atrium; RA, right atrium.

5.1.4 Catheterization and angiography

Clinical and echocardiographic features are sufficiently characteristic so that cardiac catheterization is not necessary for the diagnosis. However, cardiac catheterization is an integral part of transcatheter occlusion of the ASD.

When catheterization is performed, one will observe step-up in oxygen saturation at the right atrial level. The pulmonary venous, left atrial, left ventricular and aortic saturations are within normal range. In large defects, the pressures in both atria are equal while in small defects, an inter-atrial pressure difference is noted. The right ventricular and pulmonary arterial pressures are usually normal. Calculated pulmonary-to-systemic flow ratio (Qp:Qs) is used to quantitate the degree of shunting and a Qp:Qs in excess of 1.5:1 is considered an indication for closure of ASD.

Selective angiography in the right upper pulmonary vein at its junction with the left atrium in a left axial oblique view will reveal location and the size of the ASD. When anomalous pulmonary venous connection is suspected, selective left or right pulmonary arterial angiography should be performed and the levophase of angiogram should be scrutinized for anomalous connections.

5.1.5 Management

Despite lack of symptoms at presentation, closure of the ASD is recommended so as to 1) prevent development of pulmonary vascular obstructive disease later in life, 2) reduce chances for supra-ventricular arrhythmias and 3) prevent development of symptoms during adolescence and adulthood. Elective closure around age 4 to 5 years is recommended. Closure during infancy is not undertaken unless the infant is symptomatic. Right ventricular volume overloading by echocardiogram and a Qp:Qs >1.5 (if the child had cardiac catheterization) are indications for closure.

The conventional treatment of choice is surgical correction. While the secundum ASDs can be successfully repaired by open-heart surgical techniques with a low (<1%) mortality, the morbidity with cardiac surgery is universal, and residual scar is present in all. Because of this reason several transcatheter methods have been developed. Clinical trials have been undertaken in a large number of patients with Bard clamshell septal occluder and buttoned device and feasibility and effectiveness of these devices in occluding the ASD have been demonstrated. Fractures of one or more arms of the clamshell device with occasional embolization, has prompted the investigators and the FDA to withdraw the device from clinical trials. The buttoned device has undergone clinical trials and, immediate and short-term follow-up results are encouraging (Rao et al 1994). Recently, a large number of other devices (Das Angel-Wing, ASDOS, Amplatzer, CardioSeal, Helex and others) have been introduced and clinical trials began (Chopra and Rao 2000). However, Amplatzer and Helex are the only devices that are approved by FDA for general clinical use. The experience with Amplatzer for most defects has been encouraging. Helex device is only useful in small to medium-sized defects.

Ostium primum and sinus venosus defects are not amenable to transcatheter closure and surgical correction is the treatment of choice. In the ostium primum defect, apart from closing the ASD, the mitral valve should be repaired in such a manner as to preserve its function. In the sinus venosus defect, diversion of the anomalously connected pulmonary veins into the left atrium along with the closure of the ASD should be undertaken.

5.2 Ventricular septal defect

Ventricular septal defect (VSD) is the most common CHD and constitutes 20% to 25% of all CHDs. The defect may be small, medium or large and is classified based on its location in the inter-ventricular septum (Fyler 1992). The defect is most commonly (80%) located in the membranous septum, in the subaortic region and is commonly referred to as perimembranous defect. The defect may also be located in the conal septum in the subpulmonary region and is called supracristal defect and constitutes 5% to 7% of VSDs. This type of defect is more commonly encountered in the Far East including Japan and may constitute up to 29% of VSDs. The third type, in the posterior septum, is commonly referred to as atrioventricular canal defect and approximately 8% of the VSDs are of this type.

Finally, the defect may be located in the muscular and apical portion of the ventricular septum and may make-up 5% to 20% of all VSDs, depending on the study selected. When multiple muscular defects are seen, it is often referred to as "Swiss-cheese" type of VSD.

5.2.1 Symptoms

The clinical symptomatology is largely dependent upon the size of the VSD. In small defects, the patients are usually asymptomatic and are detected because a cardiac murmur heard on routine examination. Patients with medium and large defects may present with symptoms of congestive heart failure (dyspnea, tachypnea, sweating and failure to gain weight) or with symptoms related to bronchial obstruction and/or respiratory infection.

5.2.2 Physical findings

These, again, depend upon the size of the defect. In small defects the only abnormality is a loud holosystolic murmur (Figure 1 bottom) heard best at the left lower sternal border and is sometimes referred to as "maladie de Roger". Sometimes, the holosystolic murmur may be heard best at left mid and left upper sternal borders, depending upon the direction of the VSD jet. In very small defects, murmur, though begins with first heart sound, may not last through the entire systole; the shorter the murmur, the smaller is the defect.

In medium and large defects, the right and left ventricular impulses are increased and somewhat hyperdynamic. A thrill may be felt at the left lower sternal border. The second heart sound is split unless there is pulmonary vascular obstructive disease, in which case a loud single second heart sound is heard. The pulmonary component of the second sound may be normal or increased, depending upon the degree of elevation pulmonary artery pressure. Clicks are unusual for VSD patients although they can be heard in patients whose VSDs are undergoing spontaneous closure by aneurysmal formation of the membranous ventricular septum. A holosystolic murmur is best heard at the left lower sternal border and does not usually radiate although it may be heard widely over the precordium. The intensity of the murmur may vary between grades II-V/Vl. There is no significant variation of this murmur with respiration. This murmur is produced by flow across the VSD. The intensity of the murmur does not bear any consistent relationship with the size of the defect. A grade I-II/Vl mid-diastolic flow rumble may be heard at the apex in patients with medium to large-sized defects and large left-to-right shunts; this murmur is heard best with the bell of the stethoscope. The mid diastolic murmur is due to increased flow across the mitral valve and usually indicates a Qp:Qs greater than 2:1.

5.2.3 Noninvasive evaluation

5.2.3.1 Chest x-ray

The x-ray shows cardiomegaly and increased pulmonary vascular markings if the shunt is large. Left atrial enlargement may be noted.

5.2.3.2 Electrocardiogram

The ECG may be normal in very small defects or may show evidence for left ventricular hypertrophy in small to moderate defects while it may show biventricular or right ventricular hypertrophy in moderate to large defects. Electrocardiographic signs of left atrial enlargement may also be seen.

5.2.3.3 Echocardiogram

Echo shows increase in left atrial and left ventricular size, which is again dependent upon the size of the VSD. The location and size of the VSD can be imaged by 2-dimensional echocardiography. Left-to-right shunting across the VSD can be demonstrated by Doppler echocardiography and color mapping (Figure 10).

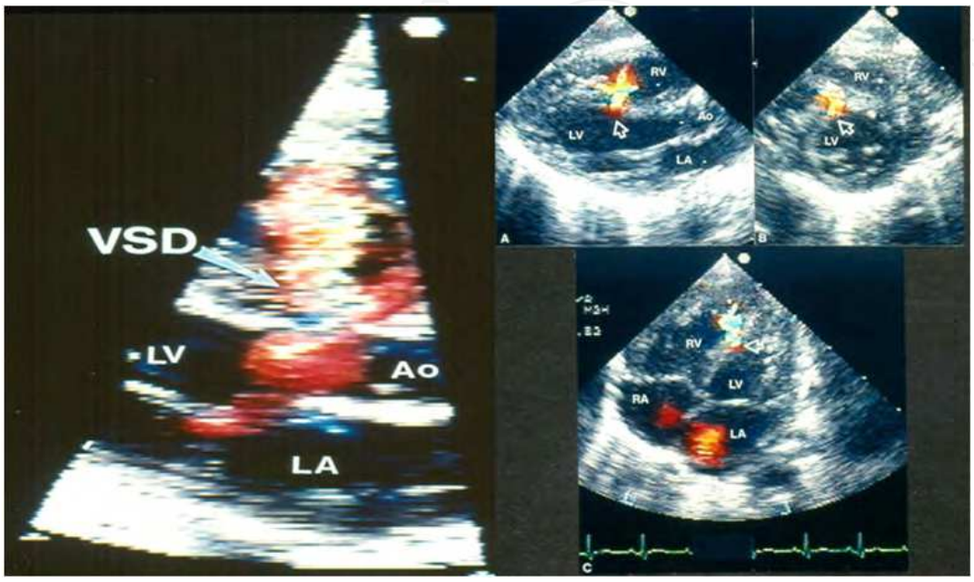

Fig. 10. Two dimensional echocardiographic views of the ventricular septum in long axis with color flow imaging (left panel) demonstrating a perimembraneous ventricular septal defect (VSD) and of the ventricular septum (arrows) in multiple views (Right panel A, B and C) with left-to-right shunt. Ao, Aorta; LA, left atrium; LV, Left ventricle; RA, right atrium; RV, Right ventricle

Peak Doppler flow velocity magnitude is inversely proportional to the size of defect. Indeed the right ventricular/pulmonary arterial pressures may be estimated by determining to peak Doppler flow velocity across the VSD.

$$\text{RV/PA peak pressure} = \text{peak arm blood pressure} - 4\,V_{VSD}^2$$

Where, RV and PA are right ventricle and pulmonary artery and V_{VSD} is the peak Doppler velocity across the VSD.

The right ventricular peak pressure may also be estimated by tricuspid back flow (regurgitant) velocity:

$$\text{RV peak pressure} = 4\,V_{TR}^2 + \text{RAP}$$

Where, V_{TR} is peak tricuspid regurgitant velocity and RAP is estimated right atrial pressure (5 mmHg).

Both formulas may help to verify internal consistency of the Doppler methodology in estimating the size of the VSD. The higher the estimated RV pressure, larger is the size of the VSD.

5.2.4 Cardiac catheterization & cineangiography

Many of the issues that require definition by catheterization in the past can be resolved by good quality echo-Doppler studies and catheterization is not routinely required. When questions cannot be satisfactorily answered, cardiac catheterization may be useful.

Step-up in oxygen saturation is observed in the right ventricle. The saturations in the left-side of the heart are usually normal. The right ventricular and pulmonary arterial pressures are normal in small VSDs and are elevated in moderate to large defects; the magnitude of elevation is proportional to the size of the VSD. Calculated Qp:Qs gives an estimate of degree of left-to-right shunting. A Qp:Qs greater than 2:1 is generally considered an indication for intervention. Pulmonary vascular resistance may be calculated:

$$PVR = (\text{Mean PA presence} - \text{Mean LA pressure})/\text{Pulmonary blood flow index}$$

Where, PVR is pulmonary vascular resistance, PA and LA are pulmonary artery and left atrium respectively.

The calculated resistance is usually 1 to 2 units and a resistance in excess of 3.0 units is considered elevated. Marked elevation of the resistance (>8.0 units) contraindicates surgical repair. When the resistance is elevated, oxygen and other vasodilating agents (Nitric oxide[NO]) should be administered to demonstrate the reversibility.

Selective left ventricular angiography in a left axial oblique view is usually required to demonstrate size and location of the VSD.

Natural history of VSDs

Knowledge of the natural history of these defects is interesting and such understanding is important in the management of children with these defects.

5.2.4.1 Spontaneous closure

Approximately 40% of VSDs spontaneously and completely close. Additional 25% to 30% of defects may become small enough not to require surgical intervention. Muscular VSDs tend to close more frequently than membraneous defects. While small defects tend to close more frequently than large defects (60% vs. 20%), even defects large enough to produce congestive heart failure or require pulmonary artery banding in infancy go on to close spontaneously. The majority of the defects close by age 2 years, most close by age 5 to 7 years, but the process of spontaneous closure continues through adolescence and adulthood. Most commonly the defect closes by apposition of leaflets of the tricuspid valve against it or by aneurysmal formation of the membranous ventricular septum.

5.2.4.2 Pulmonary vascular obstructive disease

Pulmonary vascular obstructive disease may develop in 10% of VSDs. This is probably related to the exposure of the pulmonary vascular bed to high pressure and high flow. Prompt diagnosis and closure of the defect at least prior to 18 months of age is likely to reduce the incidence of development of pulmonary vascular disease.

5.2.4.3 Development of infundibular stenosis.

Development of infundibular stenosis, the so called Gasul's transformation of the VSD may occur in 8% of the defects. There may be specific markers such as right aortic arch and increased angle of the right ventricular outflow tract that may predispose a VSD to undergo Gasul's transformation. While development of infundibular stenosis eventually requires the

patient to have surgery, it indeed protects the pulmonary vascular bed and prevents development of pulmonary vascular obstruction disease.

5.2.4.4 Aortic insufficiency

Aortic insufficiency develops in approximately 5% of patients. This may either be related to prolapse of an aortic valve cusp into the VSD or lack of support to the aortic root. This complication appears to occur more commonly with supracristal VSDs than with other types. Surgical correction is indicated if moderate to severe aortic insufficiency is present.

5.2.5 Management

The management strategies depend, to a large degree, on the size of the VSD. In small VSDs, reassurance of the parents, subacute bacterial endocarditis prophylaxis and periodic clinical follow-up are all that are necessary.

In moderate-sized defects, treatment of heart failure, if present, should be undertaken. Failure to thrive and markedly enlarged left ventricle are probably indications for surgical closure. In very large defects the heart failure should be treated aggressively. If the congestive heart failure is difficult to control with the usual anti-congestive measures or if failure to thrive is present, surgical closure should be undertaken.

In large defects with near systemic pressures in the right ventricle and pulmonary artery, surgical closure should be performed prior to 18 to 24 months of age even if heart failure control and adequate weight gain are present. Total surgical correction is currently recommended. The previously used approach of initial pulmonary artery banding in small and young babies followed by surgical closure of the VSD later is no longer recommended. However, in muscular, Swiss-cheese variety of defects, initial pulmonary artery banding may be appropriate.

When the pulmonary vascular resistance is elevated, its response to oxygen and other vasodilator agents (NO), pulmonary arterial wedge angiography and sometimes, even lung biopsy may be necessary to determine the suitability for surgical closure. Patients with calculated pulmonary vascular resistance less than 8 wood units with a Qp:Qs >1.5 are generally considered suitable candidates for surgery. If the resistance drops to levels below 8 units after administering oxygen or other vasodilator agents, the patient becomes a candidate for surgery.

Large VSDs with severe elevation of pulmonary resistance (irreversible pulmonary vascular obstructive disease) are not candidates for surgery. Symptomatic treatment and erythropheresis for symptoms of polycythemia should be undertaken. These patients may eventually become candidates for lung transplantation.

When surgery is indicated, open heart surgical technique is the treatment of choice. Several investigators have attempted transcatheter occlusion of VSD in a manner similar to ASD closure. Such methods may be feasible in muscular defects (Thanopoulos et al 1999) and membranous defects with sufficient septum in the subaortic region so that the device can be implanted without interfering with aortic valve function. Specially designed Amplatzer perimembranous VSD occluders were used to close the perimembranous VSDs in clinical trials (Fu et al 2006, Holzer et al 2006), but with significant incidence of heart block (Rao 2008). At the present, FDA has only approved Amplatzer muscular VSD occluder for transcatheter closure of muscular VSDs. Some large muscular VSDs in small babies may be

closed by hybrid procedures via sternotomy and a purse-string suture in the right ventricle under transesophageal echo guidance (Amin et al 2008). No device is yet approved for closure of perimembranous VSD, presumably because of concern for development of heart block (Rao 2008).

5.3 Patent ductus arteriosus

Ductus arteriosus, one of the fetal circulatory pathways, diverts the desaturated blood from the pulmonary artery into the descending aorta and placenta for oxygenation (Rao 1991a). After the infant is born, the ductus arteriosus constricts and closes spontaneously, presumably secondary to increased PO_2. But in some children, such spontaneous closure does not occur. This is more frequent in prematurely born infants. Patent ductus arteriosus (PDA) may be an isolated lesion and may be present in association with other defects. Isolated PDA constitutes 6 to 11% of all CHDs. In this section, isolated PDA beyond neonatal (and premature) period will be discussed. PDA is a muscular structure connecting the main pulmonary artery (at its junction with the left pulmonary artery) with the descending aorta at the level of left subclavian artery. The configuration of PDA varies considerably but most often it has a conical or funnel shape. The aortic end is wide and gradually narrows (ampulla) towards the pulmonary end. The narrowest segment is most often at the pulmonary end. Other types which are short and tubular and those with multiple constrictions and bizarre configuration can also be seen. Because of usually higher pressure and resistance in the systemic circuit than in the pulmonary circuit, left-to-right shunt takes place across the PDA. The degree of left-to-right shunting depends upon the minimal diameter of the ductus and ratio of pulmonary to systemic vascular resistance.

5.3.1 Symptoms

Clinical presentation depends upon the size of the ductus. If the PDA is small, there are no symptoms and it is usually detected because of a murmur detected on a routine examination. Moderate to large ducti with large shunt may either present with symptoms of easy fatigability, symptoms associated congestive heart failure or respiratory symptoms suggestive of lung collapse (very large ductus in small babies).

5.3.2 Physical findings

Left ventricular impulse is normal in small ducti and may be hyperdynamic with large shunts. A thrill may be felt at the left upper sternal border and in the suprasternal notch. The first heart sound is usually normal and the second heart sound may be buried within the murmur. In the majority of cases, a continuous murmur (Figure 11, top) is heard best at the left upper sternal border. The murmur begins in systole and continues through the second heart sound into the diastole. The systolic component of the murmur crescendos up to the second heart sound while the diastolic part descrescendos to a varying distance (time) into the diastole. The continuous murmur must be distinguished from the to-and-fro murmur; the latter is a combination of an ejection systolic murmur and an early diastolic descrescendo murmur (for example aortic stenosis and insufficiency) (Figure 11, bottom) (Rao 1991b).

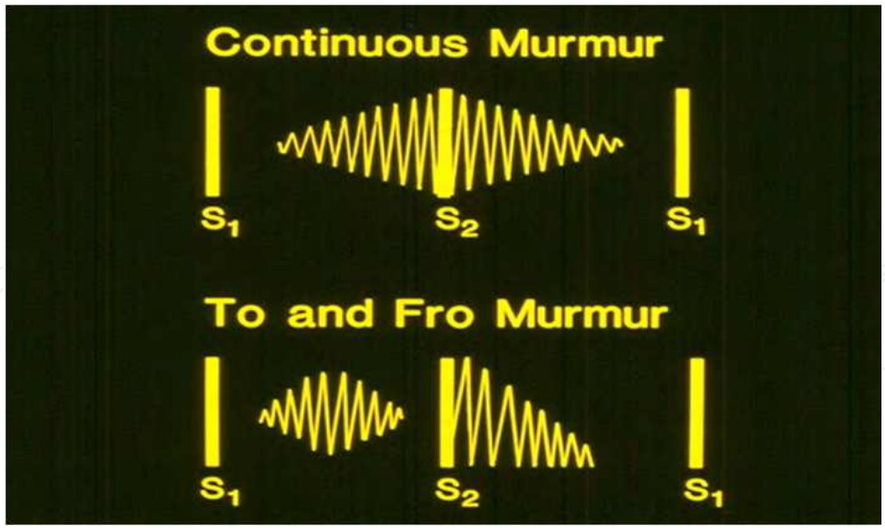

Fig. 11. Graphic representation of continuous and to-and-fro murmurs. The continuous murmur (top) begins in systole shortly after the first heart sound (S_1), crescendos up to the second heart sound (S_2) and decrescendos to a varying distance (time) into the diastole. In contradistinction to this murmur, the to-and-fro murmur (bottom) consists of an ejection systolic murmur with a separate early diastolic decrescendo murmur; note that there is a definite gap between the end of the ejection murmur and S_2.

The continuous murmur of PDA may be of grade I-V/VI in intensity. There is some beat-to-beat variation in the intensity of the murmur and for this reason it is described as machinery murmur. Multiple ejection clicks are usually heard within the murmur and this is rather characteristic of the PDA. The majority of the time, the murmur does not change with the position of the body, although the diastolic component of the murmur is heard better in a supine than in an upright position. However, in patients with very small PDA, the continuous murmur of the PDA either completely disappears or becomes only systolic in timing when the patient sits up and returns to continuous quality when the patient assumes supine position. The postulated cause of this is "kinking" of the ductus in the upright position (Thapar et al 1978). When the ductus is moderate to large in size, a mid-diastolic murmur may be heard at the apex because of increased flow across the mitral valve, such a mid-diastolic murmur suggests a Qp:Qs greater than 2:1. Arterial pulses are bounding in all but patients with very small ductus.

5.3.3 Noninvasive evaluation

5.3.3.1 Chest x-ray

Chest film may show a normal-sized heart with normal pulmonary vascular markings with small ductus while cardiomegaly, increased pulmonary blood flow and left atrial enlargement may be seen with moderate to large ductus. Collapse with secondary inflammatory process may be observed in the lung fields of small children with large ducti.

5.3.3.2 Electrocardiogram

The ECG may be normal or may show left atrial and left ventricular enlargement, depending upon the size of the ductus.

5.3.3.3 Echocardiogram

The echo may reveal varying degrees of left atrial and left ventricular enlargement, again depending upon the size of the ductus. The left ventricular contraction indices are normal unless severe myocardial dysfunction set in. Doppler echocardiography shows characteristic diastolic flow pattern in the pulmonary artery, indicative of PDA. Characteristic color flow mapping distribution is also present (Figure 12).

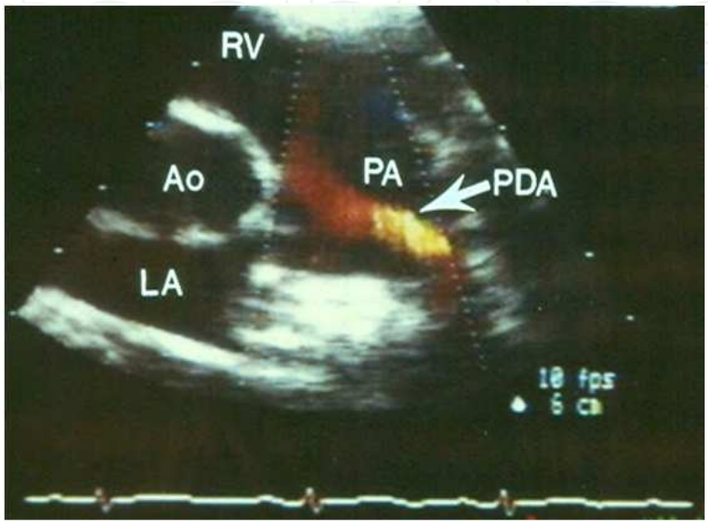

Fig. 12. Color Doppler flow mapping of the main pulmonary artery (PA) in a parasternal short axis view demonstrating the flow of the patent ductus arteriosus (PDA). Ao, Aorta; LA, Left atrium; RV, Right ventricel

5.3.4 Cardiac catheterization and selective cine angiography

These invasive studies are not necessary in the usual cases of PDA, although these procedures are integral parts of transcatheter closure.

Oxygen saturation data show a step up in oxygen saturation at the pulmonary artery level. The left heart saturations are usually normal. Calculated Qp:Qs, though usually indicates degree of shunting, it may not be accurate because of the difficulty in obtaining reliable mixed pulmonary arterial saturations. The right ventricular and pulmonary arterial pressures are normal in patients with small PDA but may be elevated if the PDA is moderate or large. Wide pulse pressure is observed in the aorta. Selective aortic arch injection demonstrates the size, shape and location of the ductus.

5.3.5 Management

It is generally believed that the presence of an isolated ductus is an indication for closure, mainly to prevent bacterial endocarditis. This can be performed at anytime, especially if associated with heart failure or pulmonary compromise. If the patient is asymptomatic, waiting until 6 to 12 months of age is generally recommended.

Until recently, surgical closure was the treatment of choice. While the risk of surgical closure is low, morbidity associated with it, namely anesthesia, endotrachial intubation and thoracotomy is universal. Because of this reason, less invasive, transcatheter closure

techniques have been developed. These transcatheter methods are increasingly being used in closing PDAs. Gianturco coil occlusion of the PDA can be performed with small caliber catheters (#4F) and is the currently preferred method of occlusion for small to medium sized ducti.

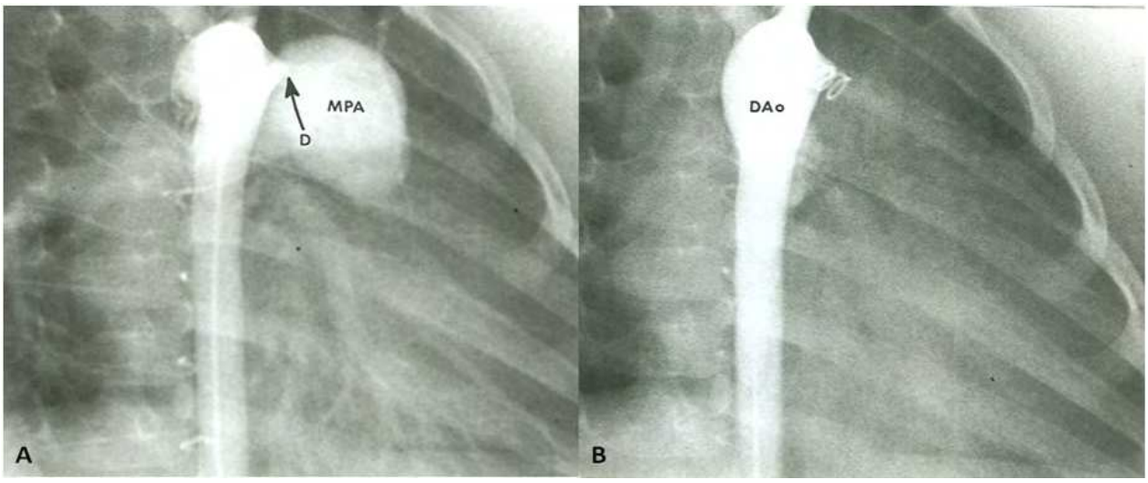

Fig. 13. Selected cine frames demonstrating a small to medium-sized patent ductus arteriosus (D) in a right anterior oblique view (A) which was occluded with a Gianturco coil (B). Dense opacification of the main pulmonary artery (MPA) prior to occlusion and no opacification following occlusion (B) are shown. DAo, descending aorta.

For large-sized PDA, surgical, video-thoracoscopic and transcatheter device closure are the available options, but most cardiologist prefer transcatheter occlusion (Rao and Sideris 1996). Amplatzer duct occluder is preferred for moderate to large PDA.

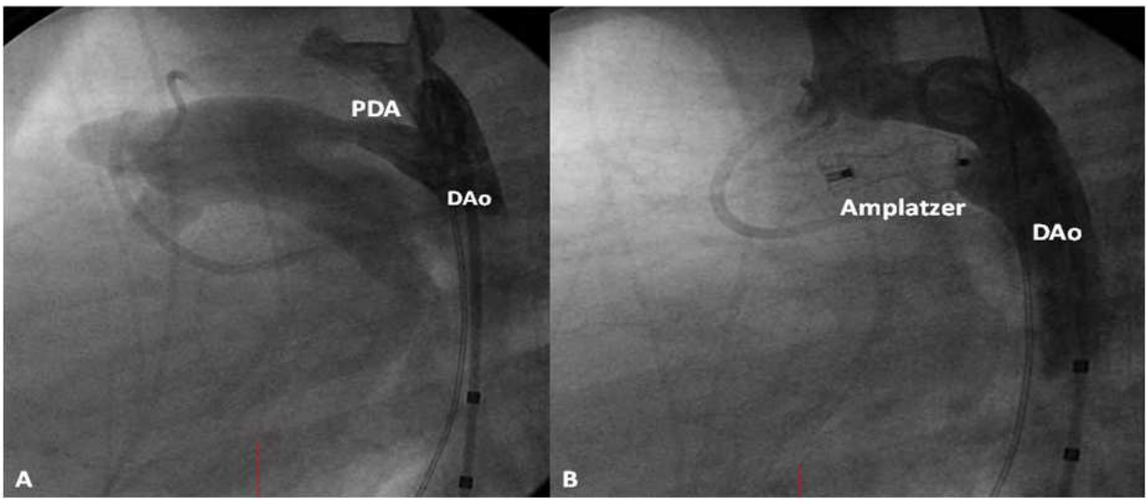

Fig. 14. Selected cine frames demonstrating a medium to large-sized patent ductus arteriosus (PDA) in a lateral view (A) which was occluded with an Amplatzer Duct Occluder (Amplatzer). Dense opacification PDA and the main pulmonary artery prior to occlusion and no opacification following occlusion (B) are shown. DAo, Descending aorta.

With wide spread use of color-Doppler echocardiography, a group of patients with color-Doppler evidence for small PDA, but without clinical features of PDA (no continuous murmur on auscultation), the so called "silent ductus" has emerged. There is no unanimity of opinion with regard to management of these patients.
Subacute bacterial endocarditis prophylaxis is recommended for all ducti prior to closure. There may not be any need for this prophylaxis three months following surgical or transcatheter closure, provided there is no residual shunt. Considerations with regard to elevated pulmonary vascular resistance with PDA are similar to those discussed under VSD section.

6. Right-to-left shunts (cyanotic heart defects)

In cyanotic congenital heart defects systemic venous blood bypasses the pulmonary circulation and gets shunted across into the left side of the heart. Thus, there is systemic arterial desaturation. By definition, cyanotic congenital heart disease does not include cyanosis due to intrapulmonary right-to-left shunting and pulmonary venous desaturation secondary to congestive heart failure. There are usually multiple defects of the heart causing right-to-left shunt. Obstruction to pulmonary blood flow (for example tetralogy of Fallot), complete admixture of pulmonary and systemic venous returns (for example, total anomalous pulmonary venous return and double-inlet left ventricle) and parallel rather than in-series circulation (transposition of the great arteries) are the causes of right-to-left shunts and cyanosis. The most important of the cyanotic CHDs are what are called "5 Ts" and are listed in table 2.

1. Tetralogy of Fallot
2. Transposition of the great arteries
3. Tricuspid atresia
4. Total anomalous pulmonary venous connection
5. Truncus arteriosus

Table 2. Common Cyanotic Congenital Heart Defects (5 Ts)

Three of these defects, namely tetralogy of Fallot, transposition of the great arteries and tricuspid atresia will be reviewed in this chapter.

6.1 Tetralogy of Fallot

Tetralogy of Fallot (TOF) is the most common cause of cyanosis beyond one year of age and constitutes 10% of all congenital heart defects. Fallot defined it as a constellation of four abnormalities to include a VSD, PS, right ventricular hypertrophy and dextroposition of the aorta. The ventricular defect is always large and non-restrictive and is located in the membranous septum in the subaortic region. Pulmonary stenosis is variable in severity and nature of obstruction. The right ventricular outflow obstruction may be mild resulting in initial left-to-right shunt at ventricular level or it may be severe causing severe cyanosis

even in the neonatal period. It may be completely obstructed (pulmonary atresia) so that there is no forward flow from the right ventricle into the pulmonary artery, thus ductal dependent. The obstruction may be infundibular, valvar or supravalvar in nature or may involve branch pulmonary arteries. The stenotic component may be at a single site or may involve multiple sites. Infundibular obstruction is the most common obstruction in TOF and is due to malposition of the crista supraventricularis. The valvar stenosis may be due to valve leaflet fusion or due to valve ring hypoplasia. Right ventricular hypertrophy of severe degree is present in all cases. Dextroposition or over-riding of aorta over the ventricular septum is a variable phenomenon. The aorta is large and is thought to be due to a developmental anomaly rather than the result of physiologic abnormality of TOF. Right aortic arch is present in 25% of TOF cases.

Atrial septal defects may be present in 15% of patients with TOF in which case it may be called pentology of Fallot. Coronary artery anomalies are present in a small but significant number of cases. Origin of the left anterior descending coronary artery from the right coronary artery is the most common coronary anomaly in TOF and sometime the course of the coronary artery may be intra-myocardial.

Because the VSD is large, the systolic pressures in both ventricles are equal and for practical purposes both ventricles act as one functional chamber. The quantity of blood flow into to the systemic and pulmonary circuits depends upon their respective resistances. The level of systemic vascular resistance and the resistance offered by the right ventricular outflow tract stenosis determine the flows. The more severe the PS, the less is the pulmonary flow. In the average case of tetralogy of Fallot, the resistance offered by PS is more than that of the systemic vascular tone with consequent right-to-left shunt across the VSD. The consequent cyanosis and hypoxemia stimulate bone marrow (via kidney and erythropoitin) and produce polycythemia. While the polycythemia is helpful in increasing oxygen carrying capacity, it becomes counter-productive when the hematocrit is excessive (60 to 70%).

6.1.1 Symptoms

The clinical presentation depends upon the degree of PS. With milder degrees of PS, symptoms may not be present until late childhood while with severe PS the presentation may be in the early infancy. Typically the infant may be pink (not cyanotic) as a neonate and develops cyanosis between 2 to 6 months of age. Most usual modes of presentation are asymptomatic murmur discovered on routine auscultation, bluish color (cyanosis) observed by the parent or primary physician, hypercyanotic spells, and decreased exercise tolerance.

Hypercyanotic spells are variously described as anoxic spells, hypoxic spells, blue spells, paroxysmal dyspnea, paroxysmal hyperpnea and so no. The spells characteristically occur in tetralogy although they can be present in other lesions with similar physiology. They can occur any time between 1 month and 12 years of age but the peak incidence is 2 to 3 months. They can occur at any time of the day but most commonly seen after awakening from sleep; crying, defecation and feeding are the common precipitating factors. Spells are characterized by increasing rate and depth of respiration (hyperpnea) with increasing cyanosis, progressing to limpness and syncope, occasionally terminating in convolutions, cerebrovascular accident or death. Spells may occur in tetralogy with mild arterial desaturation and conversely may not be present in patients with severe cyanosis.

The cause or mechanism of onset of spells is not clear. Right ventricular infundibular spasm, precipitated by acute increase in endogenous catacholamines has been proposed as a

mechanism. Prevention of these spells by beta-adrenergic blockade may further support this hypothesis. Since the spells have also been observed in patients with VSD and pulmonary atresia in whom infundibular spasm is singularly irrelevant, it is unlikely that the infundibular spasm is the cause in all cases. Another mechanism proposed is paroxysmal hyperpnea. During sleep oxygen consumption is reduced and there is a normal acid base balance. When the infant awakens the O_2 consumption increases and there is a slight acid base imbalance. There are adjustments made by the respiratory center to bring the imbalance back to normal. But, if there is a sudden increase in activity and consequent increase in oxygen consumption before the adjustments occur, decrease in PO_2 and pH and increase in PCO_2 take place triggering a hyperpnea response from the respiratory center and enter a vicious cycle. Hyperpnea reduces mean intrathoracic pressure, which decreases systemic and pulmonary resistances. Decreased systemic resistance is not matched with increased pulmonary flow because of dominant right ventricular outflow tract obstruction. Thus, there is even greater right-to-left shunt, further decreasing the PO_2 and pH and thus a vicious cycle (Guntheroth et al 1965, Rao 1983). Most workers believe that this is the most likely mechanism for the development of spells.

6.1.2 Physical examination

Central cyanosis is observed in most cases of' tetralogy of Fallot. However, it should be noted that mild arterial desaturation may not cause clinically detectable cyanosis. Clubbing of fingers and toes is observed beyond the first few months of life. There are usually no signs of congestive heart failure. Prominent right ventricular impulse or heave may be present. A systolic thrill may be present at the left upper sternal border. The first heart sound may be normal or slightly increased. The second heart sound is single without an audible pulmonary component. A grade III-IV/Vl long ejection systolic murmur, caused by blood flow through the right ventricular outflow tract, is usually heard at the left upper sternal border. In contrast to PS with intact ventricular septum, the murmur of tetralogy becomes shorter and less intense with increasing severity of PS. During hypercyanotic spell the murmur disappears or becomes very soft. A holosystolic murmur of VSD may be heard at the left lower sternal border in some children especially in less severe and acyanotic forms of tetralogy of Fallot. Early diastolic murmurs do not occur with TOF; the exception is TOF with syndrome of absent pulmonary valve. Continuous murmur of associated PDA is rarely heard. Older children may have an audible continuous murmur of bronchial collateral flow into the lungs.

6.1.3 Noninvasive evaluation

6.1.3.1 Chest roentgenogram

On a chest roentgenogram the heart size is usually normal to minimally increased. An uplifted apex, thought to indicate right ventricular hypertrophy may be present and is described by some as "boot-shaped" heart. Concavity in the region of pulmonary conus, reflecting hypoplasia of the pulmonary outflow tract may be present. Pulmonary vascular markings are usually diminished. A right sided aortic arch may be present. While a right aortic arch is expected to be present in 25% of TOF patients, the presence of a right aortic arch along with concave pulmonary conus and decreased pulmonary vascular markings in a chest x-ray makes the diagnosis of TOF virtually certain.

6.1.3.2 Electrocardiogram

The ECG shows signs of right ventricular hypertrophy. Right atrial enlargement is less commonly seen.

6.1.3.3 Blood work

Hemoglobin and hematocrit along with red blood cell indices should be monitored periodically in all children with cyanotic congenital heart defects including TOF. The degree and duration of hypoxemia determine the level of hemoglobin. In the absence of adequate iron intake, relative anemia with hypochromia and microcytosis may develop. Because this is a risk factor for developing cerebrovascular accidents, the relative anemia should be treated with oral supplemental iron.

6.1.3.4 Echocardiogram

The echo is very helpful in confirming the diagnosis and in evaluating several of the issues related to TOF. Enlargement of the right ventricle, large VSD, aortic over-ride and right ventricular outflow tract obstruction can be imaged. Shunting across the VSD and increased Doppler flow velocity across the right ventricular outflow tract can be demonstrated. Size of the main and proximal branch pulmonary arteries can be evaluated although the distal pulmonary arteries cannot easily be seen by echocardiogram.

6.1.4 Cardiac catheterization and angiography

Oxygen saturation data reveal systemic venous and arterial desaturation, usually proportional to the degree of right ventricular outflow obstruction. Usually no left-to-right shunts are demonstrated. Pulmonary venous and left atrial saturations are usually normal. The left ventricular and aortic saturations are diminished because of right-to-left shunt across the VSD. Aortic saturation is a better (than left ventricular) indicator of the degree of desaturation because of better mixing distally. The peak systolic pressures in both ventricles are equal because of a large VSD. The top of the right ventricular pressure curve is flat when compared to that of patients with PS with intact ventricular septum in which it is triangular. The pulmonary arterial pressures are low to normal with demonstrable peak systolic gradients across the pulmonary valve and infundibulum. However, multiple gradients may not be demonstrable in all patients either because of technical (multiple holes in the catheter or rapid withdrawal) or physiologic reasons. Angiographic data should be used to supplement pressure information for assessment of degree and level of right ventricular outflow obstruction. The left ventricular and aortic pressures are normal without any gradient across the aortic valve.

Angiographic evaluation of anatomy of TOF is generally recommended prior to total surgical correction, although at some centers detailed echocardiographic data may be considered adequate. Selective left ventricular angiography in a left axial oblique view to demonstrate the size and function of the left ventricle and the size and location of the VSD, particularly to exclude muscular VSD is important. Similarly selective right ventricular angiography to study its architecture, size and function and to evaluate right ventricular outflow obstruction is recommended. Pulmonary arteriogram in a sitting up view to visualize the size of the main and branch pulmonary arteries and to exclude branch pulmonary artery stenosis should be obtained. Aortic root angiography is also necessary to visualize coronary artery anatomy, especially to exclude coronary arteries crossing the right ventricular infundibulum. Origin of the left anterior descending coronary artery from the

right coronary artery occurs in a significant number of cases of TOF and should be excluded, if necessary, by selective coronary angiography.

Management

The goal of management of TOF patients is to allow total surgical correction with minimal mortality and morbidity and to prevent or treat complications inherent to cyanotic heart defects in general and TOF in particular. Protection against subacute bacterial endocarditis, prevention and/or prompt treatment of dehydration, and periodic monitoring for relative anemia secondary to iron deficiency and prompt treatment when found should be undertaken. Palliative or corrective surgical procedures should be performed prior to development of significant polycythemia. Exercise, as tolerated should be permitted unless symptoms develop with activity.

Treatment of an infant with cyanotic spell may be summarized (Rao 1989a) as follows:

1. The infant should be placed in a knee-chest position. The reason for its effectiveness appears to be related to its effect in increasing the systemic vascular resistance and thus decreasing the right-to-left shunt and improving the pulmonary flow.
2. Humidified oxygen via a facemask should be administered. Since the major defect in the spell syndrome is pulmonary oligemia rather than alveolar hypoxia, oxygen administration has limited usefulness. If the infant is unduly disturbed by the facemask, oxygen therapy may be discontinued.
3. Morphine sulfate, 0.1 mg/kg subcutaneously, may be effective in aborting the spell. The mechanism of action is not clearly delineated, but its depressive effect on the central nervous system respiratory drive (thus reducing hyperpnea) and sedation of the infant may be important.
4. Once the physical examination is completed (and the limited but important laboratory studies are obtained) the infant should be left undisturbed and allowed to rest; this in itself may improve the infant's condition.
5. Correction of metabolic acidosis (with sodium bicarbonate), anemia (by blood transfusion), and dehydration (by appropriate fluids), if present, is very important at this stage.
6. If the spell continues, vasopressors to increase the systemic vascular resistance and thus increase the pulmonary blood flow may be tried. In our experience, methoxamine (Vasoxyl) an alpha agonist has been most helpful. It is a pure peripheral vascular stimulator without any direct action on the heart. Methoxamine 20-40 mg in 250 ml of 5% dextrose in water may be administered intravenously; the rate of infusion should be adjusted to increase the systolic blood pressure by 15 to 20% of the control value. Instead, phenylephrine may be given to increase systemic vascular resistance.
7. Alternatively, propranolol, 0.1 mg/kg body weight, diluted in 50 ml of 5% dextrose in water, may be slowly administered intravenously while monitoring the heart rate (by ECG if possible). Should there be marked bradycardia, propranolol should be stopped. Once it is found to be effective, the infant may be switched to oral propranolol 1-4 mg/kg/day in three and four divided doses. The mechanism of action of propranolol is not clearly understood, but may include negative inotropic effect on the right ventricular infundibular myocardium, prevention of decrease in systemic vascular resistance and/or prevention of ventilatory response (hyperpnea) to hypoxia, all through beta adrenergic blockade. Esmolol, a rapid acting beta blocker, may also be

used. The recommended loading dose of Esmolol is 500 mcg/kg followed by 50-100 mcg/kg/min.

8. Infrequently, general anesthesia may be necessary to break the spell.
9. If the infant does not improve with any of the aforementioned measures, an emergency systemic-to-pulmonary artery shunt (the author prefers modified Blalock-Taussig anastomosis) should be performed. Occasionally, total correction, if the anatomy is adequate, may he performed on an emergency basis. The important principle is that the infant requires more pulmonary blood flow.

If the infant improves with the management outlined above, total surgical correction of the cardiac defects, if anatomically feasible, or a systemic-to-pulmonary artery shunt to improve pulmonary blood flow on an elective basis within the next day or so may be performed. More recently, we have used balloon pulmonary valvuloplasty as an alternative to Blalock-Taussig shunt, especially if valvar obstruction is a significant component of right ventricular outflow obstruction (Rao et al 1992). Another alternative to surgery is oral propranolol (dosage as above) which may help postpone surgery by several months to years.

Total surgical correction to include closure of VSD in such a manner as to direct left ventricular output into the aorta and resection of the infundibulum and/or relief of pulmonary valvar obstruction can be performed almost at any age. Enlargement of the right ventricular outflow tract with a pericardial patch (or other prosthetic material) may be necessary in some cases. Sometimes total corrective procedures are not feasible with "respectable" mortality either because of pulmonary arterial (and/or annular) hypoplasia, "smallish" left ventricle, and/or anomalous course of a major coronary artery in the right ventricular infundibulum. Size and age of the patients also enter into such decision making. If it is deemed that a given patient is not suitable for total surgical correction, palliative surgery may be utilized to augment pulmonary blood flow and to allow the patients to grow into an age, size and anatomy that are more likely suitable for complete correction. Classic or modified Blalock-Taussig shunt is clearly a preferred surgical method for this purpose. We have used balloon pulmonary valvuloplasty in TOF patients to augment pulmonary blood flow and to allow for growth and development of the pulmonary arterial system and left ventricle so that a total surgical corrective procedure could be performed at a later time with a greater chance for success (Rao et al 1992).

Discussion of the management of TOF with pulmonary atresia, TOF with MAPCAs (multiple aorto-pulmonary collateral arteries) and TOF with syndrome of absent pulmonary valve is beyond the scope of this chapter and the reader is referred elsewhere (Alapati and Rao 2011) or to the standard textbooks.

6.2 Transposition of the great arteries

Transposition of the great arteries (TGA) is the most common cyanotic congenital heart defect presenting in the newborn period. It constitutes 5% of all CHD and 10% of all cyanotic CHD. There are multiple definitions used to describe TGA. Perhaps, the most accurate description is “a condition in which the aorta arises from the morphologic right ventricle and the pulmonary artery from the morphologic left ventricle”. In the most common form, usually referred to as complete transposition, the atria are normal in position (situs solitus of the atria), there is atrioventricular concordance (right atrium connected to the right ventricle and the left atrium to the left ventricle), d loop of the ventricles (right ventricle on the right and left ventricle on the left), and ventriculo-arterial discordance (aorta

arising from the right ventricle and the pulmonary artery from the ventricle). The systemic venous blood from the vena cavae enters the right atrium and right ventricle and from there into the aorta while the pulmonary venous blood enters the left atrium and left ventricle and from there into the pulmonary artery. Thus, the circulation is parallel instead of normal in-series circulation. Because of this reason, the systemic venous blood does not get oxygenated and the pulmonary venous blood does not get delivered to the body. The infant will not survive unless there are intercirculatory shunts such as atrial or ventricular septal defect or patent ductus arteriosus.

6.2.1 Symptoms

Clinical features depend upon the anatomic type, namely Group I, TGA with intact ventricular septum; Group II, TGA with VSD, and Group III, TGA with VSD and PS (Rao 2010).

In group I with intact septum, the infants usually present with cyanosis within the first week of life. They may otherwise be asymptomatic. However, they will soon become tachypnoeic and develop respiratory distress. If they are not appropriately treated, they become acidotic and go on to become lethargic without lack of spontaneous movement, and eventually die.

Group II TGA patients with VSD present with symptoms of congestive heart failure (tachypnea, tachycardia, sweating, and poor feeding) between 4 to 8 weeks of life, but the cyanosis is minimal.

Group III patients (TGA with VSD and PS) have variable presentation, depending upon the severity of PS. If there is poor mixing, they may present early in life and mimic TGA with intact septum. If the PS is severe, the presentation is essentially similar to that described in the TOF section. With moderate PS the presentation is late with longer survival. With mild PS, congestive heart failure signs may be present, similar to group II patients.

6.2.2 Physical examination

The group I patients with intact septum are usually severely cyanotic but are without distress until severe hypoxemia and acidosis develop. Clubbing is not present in the newborn period and may not develop until 3 to 6 months. The right ventricular impulse is increased and the second heart sound is single. Either no murmur or a grade I-II/Vl nonspecific ejection systolic murmur may be auscultated. In group II patients, tachypnea, tachycardia, minimal cyanosis, hepatomegaly, increased right and left ventricular impulses, single second sound, a grade III-IV/VI holosystolic murmur at the left lower sternal border and a mid-diastolic flow rumble (murmur) at the apex may be present. In group III patients, the findings are similar to TGA with intact septum, TGA with VSD, or TOF depending upon the degree of mixing and severity of PS.

6.2.3 Noninvasive evaluation

6.2.3.1 Chest roentgenogram

Chest x-ray in the intact septum group is benign with normal to minimal cardiomegaly and normal to slightly increased pulmonary vascular marking. The thymic shadow may rapidly involute and a narrow pedicle (superior mediastinum) may be seen. A combination of the above signs may sometimes assume "egg-shaped" appearance on a postero-anterior chest

roentgenogram. In group II patients with VSD, moderate to severe cardiomegaly and increased pulmonary vascular markings are usually seen. In group III patient, mild to at worst moderate cardiomegaly may be observed. The pulmonary vascular marking may be increased, normal or decreased, dependent upon the severity of PS.

6.2.3.2 Electrocardiogram

The ECG in a neonate with TGA and intact septum (Group I) may be normal with the usual right ventricular preponderance seen during this age. In older infants clear-cut right ventricular hypertrophy is seen and in addition right atrial enlargement may be observed. In group II patients, biventricular hypertrophy and left atrial enlargement are usual. In group III, right ventricular or biventricular enlargement is seen.

6.2.3.3 Echocardiogram

The echo is usually helpful in the diagnosis and assessment. Demonstration of transposition of the great arteries is somewhat difficult in view of the fact that atrial and ventricular anatomy is normal and the aortic and pulmonary valves look similar on echocardiographic study. If one can follow the great vessel arising from the left ventricle and demonstrate its bifurcation, identifying it as a pulmonary artery, the diagnosis is easy. One of the helpful indirect signs is somewhat a posterior course the great vessel off of the left ventricle in a precordial long axis view, indicating pulmonary artery in contradistinction to anteriorly coursing ascending aorta. On-end visualization of the aorta and pulmonary artery on a precordial short axis view of the heart is also helpful in suggesting TGA. The presence of an inter-atrial communication and patent ductus arteriosus and shunt across them by color and pulsed Doppler can also be evaluated. In addition to these, demonstration of VSD and PS will place the patients into the respective groups.

6.2.3.4 Other laboratory data

Blood gas values are useful in demonstrating the degree of hypoxemia and ventilatory status. Hemoglobin and hematocrit are particularly useful in the follow-up of older children.

6.2.4 Cardiac catheterization and angiography

With the increasing accuracy of echocardiographic diagnosis, invasive studies are not necessary for diagnosing TGA. Need for rapid relief hypoxemia and acidosis by balloon atrial septostomy and the need for a greater definition of coronary artery anatomy prior to arterial switch procedure may necessitate catheterization and angiography.

In group I patients, vena caval, right atrial, right ventricular and aortic saturations are moderately to severely diminished unless atrial, ventricular or ductal shunting is present. Similarly, the pulmonary venous, left atrial, left ventricular and pulmonary arterial saturations are high with minimal, if any right-to-left shunt. In TGA, the pulmonary artery saturations are higher than those in the aorta.

The left atrial pressure is usually high with a pressure gradient across the atrial septum. The right ventricular pressure is at systemic level without any gradient across the aortic valve. In TGA with intact septum the left ventricular and pulmonary artery pressure are normal without any gradient across the pulmonary valve. However, in the early neonatal period, prior to involution of the pulmonary vasculature, these pressures are elevated, compared to normal. In the presence of significant VSD and/or PS, the left ventricular pressure is elevated and this is usually proportional to the size of VSD and severity of PS. The

pulmonary artery pressure is usually increased with associated VSD while with PS it may be low to normal.

Selective right ventricular angiography reveals a morphologically right ventricle with opacification of an anteriorly and superiorly displaced aorta. The aortic valve is located to the right of the pulmonary valve (d-TGA). The aorta ascends in a normal fashion and usually descends on the left side of the spine. The size and function of the right ventricle and presence of tricuspid insufficiency should be evaluated. If a VSD is present, it may be visualized. A laid-back view of the aortic root angiography along with a lateral view may be useful in demonstrating coronary artery anatomy. Aortography may, in addition, be useful in demonstrating PDA and CoA. Left ventricular cineangiogram reveals a morphologic left ventricle with prompt opacification of the pulmonary artery. The pulmonary valve is located posterior, inferior and to the left of the aortic valve. Left ventricular angiography should be scrutinized for subvalvar and valvar PS. A VSD may be visualized, if present.

6.2.5 Management

Untreated, TGA with intact septum carries a poor prognosis. The initial management of this and other cyanotic neonates is similar. Monitoring the infant's temperature and maintenance of neutral thermal environment is extremely important. In hypoxemic infants, ambient oxygen should be administered. In cyanotic CHD patients, no more than 0.4 FIO_2 is necessary. Metabolic acidosis (pH < 7.25), if any, should be corrected with sodium bicarbonate (usually 1-2 mEq/kg diluted half and half with 5% or 10% dextrose solution) immediately. Respiratory acidosis should be cared for by appropriate suctioning, intubation and assisted ventilation. Hypoglycemia may be a significant problem; therefore, the infant's serum glucose should be monitored and the neonates should routinely receive 10% dextrose in water intravenously. If hypoglycemia (<30 mg/100ml) occurs, 15% to 20% dextrose solution should be administered. Similarly hypocalcemia should be monitored for and treated, if found. If an infant is getting progressively hypoxemic, it is likely that the intercirculatory pathways (patent foramen ovale and patent ductus arteriosus) are closing. Prostaglandin E_I (PGE_I) (0.05 to 0.1 mcg/kg/min) intravenously may help open the ductus, thus improve oxygenation. Balloon atrial septostomy may be necessary to improve hypoxemia even after PGE_1. Total surgical correction by arterial switch procedure (Jatene) is the treatment of choice in these neonates and will be discussed here-under.

TGA patients with VSD usually present with heart failure and aggressive anticongestive measures are indeed needed. Balloon atrial septostomy may help relieve pulmonary venous congestion and improve oxygenation. These patients will require Jatene procedure with closure of VSD.

TGA patients with VSD and PS may have varying presentation. If the reason for hypoxemia is poor mixing, balloon atrial septostomy is the treatment of choice. If the hypoxemia is secondary to decreased pulmonary flow, a Blalock-Taussig type of shunt may be needed. Sometimes both balloon septostomy and balloon dilatation of pulmonary valve may be performed via catheters in some of these children. Eventually these patients require a Rastelli type of repair.

Mustard procedure, which was originally described in 1964 was the most commonly used operation for TGA in the past. In this operation, hemodynamic correction of the defect is achieved by re-directing the systemic and pulmonary venous returns by means of an intra-atrial baffle. Better understanding of the conduction system and its blood supply coupled

with the use of a pericardial baffle (instead of Dacron baffle) has significantly reduced post-operative complications such as arrhythmia and baffle obstruction. Other types of atrial switch operations, originally described by Senning and by Shumacker have also been used in several centers. When venous switch procedure is opted for, Mustard and Senning procedures appear to be selected with equal frequency, depending upon the institution/surgeon. In 1975, Jatene described anatomical corrections for TGA by arterial switch with relocation of the coronary arteries. Initially this procedure was performed for TGA with non-restrictive VSD and subsequently was adapted to TGA with intact septum. The arterial switch procedure has several advantages over the venous switch procedure in that the arrhythmias are less frequent, and the left ventricle rather than the right ventricle serves as a pump to systemic circuit. Arterial switch procedure, however, must be performed in the early neonate prior to deconditioning the left (pulmonary) ventricle in TGA patients with intact septum. Although there are no extensive long term follow-up results available, the short term and medium-term follow-up results are very encouraging and, at this time, the arterial switch procedure with or without LeCompte maneuver is considered the preferable operation for patients with TGA.

6.3 Tricuspid atresia

Tricuspid atresia (TA) is a cyanotic, congenital cardiac anomaly and has been commonly defined as congenital absence or agenesis of the morphologic tricuspid valve. It is the third most common cyanotic CHD and is the most common cause of cyanosis with left ventricular hypertrophy. Tricuspid atresia accounts for 1.4% of subjects with CHD. The most common type of TA, muscular variety, is characterized by a dimple or a localized fibrous thickening in the floor of the right atrium at the expected site of the tricuspid valve. The right atrium is usually enlarged and its wall thickened and hypertrophied. An inter-atrial communication, which is necessary for survival, is usually a stretched patent foramen ovale. The left atrium is enlarged and may be more so if the pulmonary blood flow is increased. The mitral valve is morphologically a mitral valve, usually bicuspid but its orifice is large and rarely incompetent. The left ventricle is clearly a morphologic left ventricle with only occasional abnormality; however, it is enlarged and hypertrophied. The VSD may be large, small or non-existent (intact ventricular septum) or multiple VSDs may be present. While a variety of VSDs are seen in TA hearts, muscular defects are most common. Also, most of these VSDs are restrictive and produce subpulmonary stenosis in patients with normally related great arteries and subaortic stenosis in patients with transposed great arteries. The right ventricle is small and hypoplastic; its size, by and large, is determined by the anatomic type. The relative position of the great vessels is quite variable and has been the basis for classification of' this anomaly: Type I, normally related great arteries; Type II, d-transposition of the great arteries; Type III, malpositions of the great arteries other than d-transposition; and Type IV, Truncus arteriosus (Table III) (Rao 1980).

Pulmonary outflow tract obstruction may be either subvalvar or valvar in patients with transposition while in patients with normally related great arteries, it is often at the VSD level although, in a few cases, subvalvar pulmonary stenosis, narrow tract of the hypoplastic right ventricle and, rarely, valvar PS may also be responsible for pulmonary outflow tract obstruction. The pulmonary artery may be atretic and in such cases a PDA or aorto-pulmonary collateral vessels may be present. Association with aortic coarctation is rare with type I patients and is more common in patients with transposition of the great arteries.

An obligatory right-to-left shunt occurs it the atrial level in most types and subtypes of TA. Thus, the systemic and coronary venous blood mixes with pulmonary venous return in the left atrium and exits into the left ventricle. In type I (normally related great arteries) patients with a VSD, left-to-right ventricular shunt occurs, thus perfusing the lungs. In the absence of a VSD (i.e., intact ventricular septum), the pulmonary circulation is derived either via a PDA or through aorto-pulmonary collateral vessels. The presence of either a VSD or other means of blood supply to the lungs is essential for the patient's survival. The aortic blood flow is derived directly from the left ventricle. In type II (with d-transposition of the great arteries), the pulmonary blood flow is directly divided from the left ventricle. The systemic blood flow is via the VSD and right ventricle.

Type I	Normally related great arteries	Each Type and Subtype are divided
Type II	D-transposition of the great arteries	Subgroup a. Pulmonary atresia
Type III	Malpositions of the great arteries other than D-transposition	Subgroup b. Pulmonary stenosis or hypoplasia
		Subgroup c. Normal pulmonary arteries (no pulmonary stenosis)
	Subtype 1. L-transposition of the great arteries	
	Subtype 2. Double outlet right ventricle	
	Subtype 3. Double outlet left ventricle	
	Subtype 4. D-malposition of the great arteries (anatomically corrected malposition)	
	Subtype 5. L-malposition of the great arteries (anatomically corrected malposition)	
Type IV	Persistent truncus arteriosus	

Table 3. A Unified Classification of Tricuspid Atresia

6.3.1 Symptoms

Approximately one-half of the patients with TA present with symptoms on the first day of life and 80% would have had symptoms by the end of the first month of life. The magnitude of pulmonary blood flow determines the timing of and, type of clinical presentation. Two modes of presentation are recognized; those with decreased pulmonary blood flow and those with increased pulmonary blood flow.

Infants with pulmonary oligemia present with symptoms of cyanosis within the first few days of life; the more severe the pulmonary oligemia, the earlier is the clinical presentation. These hypoxemic infants may have hyperpnea and acidosis if the pulmonary blood flow is markedly diminished. The majority of the infants belong to type Ib (no transposition and pulmonary hypoplasia with a small VSD). Patients with pulmonary atresia (subgroup a, of all types) irrespective of great vessel relationship will also present with early cyanosis, especially when the ductus begins to close. Hypoxic spells are not common in the neonate although the spells can occur later in infancy.

Infants with pulmonary plethora usually present with signs of heart failure within the first few weeks of life although an occasional infant may present within the first few days of life. They are only minimally cyanotic, but present with symptoms of dyspnea, fatigue, difficulty to feed, and perspiration. Recurrent respiratory tract infections and failure to thrive are other modes of presentation. The majority of these patients belong to type II (transposition with a large VSD) although a small number of patients may be of type Ic (no transposition but a large VSD). The association of aortic coarctation with type II patients has already been mentioned and coarctation, when present, makes them vulnerable to early cardiac failure.

6.3.2 Physical findings

In infants (and children) with pulmonary oligemia, physical examination reveals central cyanosis, clubbing (in older infants and children), tachypnea or hyperpnea, normal pulses, prominent "a" wave in the jugular venous pulse (if there is inter-atrial obstruction), and no hepatic enlargement. Quiet precordium, and absence of thrills are usual. The second heart sound is usually single. A holosystolic murmur suggestive of VSD may be heard at the left lower or mid sternal border. No diastolic murmurs are heard. In patients with associated pulmonary atresia, no murmurs are usually heard, although in an occasional patient a continuous murmur of PDA may be heard. Signs of clinical congestive heart failure are notably absent.

In the group with pulmonary plethora, examination reveals tachypnea, tachycardia, decreased femoral pulses (if associated with CoA), minimal cyanosis and hepatomegaly. Prominent "a" waves in the jugular veins and/or presystolic hepatic pulsations may be observed with associated inter-atrial obstruction. The second heart sound may be single or split. A holosystolic murmur of VSD is usually heard at the left lower sternal border. An apical mid-diastolic flow murmur may be heard. Clear-cut signs of congestive heart failure are usually present.

6.3.3 Non-invasive evaluation

6.3.3.1 Chest x-ray

Chest film appearance is, by and large, dependent upon the total pulmonary blood flow. In patients with diminished pulmonary blood flow (the majority of infants fall into this group), the heart size is either normal or minimally enlarged. Several patterns of cardiac configuration have been described but in the author's experience and that of others, there is no consistent pattern that would be diagnostic of TA. There may be concavity in the region of pulmonary artery segment in patients with pulmonary oligemia and small pulmonary artery. The right atrial shadow may be prominent. In patients with increased pulmonary blood flow, cardiomegaly and prominent pulmonary vasculature are seen.

6.3.3.2 Electrocardiogram

The ECG can be virtually diagnostic of tricuspid atresia in an infant with cyanotic CHD. The characteristic features include right atrial enlargement, an abnormal, superiorly oriented major QRS vector (so called left axis deviation) in the frontal plane, left ventricular hypertrophy and diminished right ventricular forces. Abnormally superior vector (left axis deviation) is present in excess of 80% of patients with type I (normally related great vessels) anatomy while only less than 50% of patients with type II (transposition) anatomy show such a typical electrocardiographic pattern.

6.3.3.3 Echocardiogram

The echo is reasonably characteristic for TA. Two-dimensional echocardiography, apart from showing enlarged right atrium, left atrium, and left ventricle and a small right ventricle, will demonstrate the atretic tricuspid valve directly. In the most common muscular type, a dense band of echoes is seen at the site where the tricuspid valve should be and the anterior leaflet of the detectable atrioventricular valve is attached to the left side of the inter-atrial septum. Apical and subcostal four-chambered views are best to demonstrate the anatomy. Atrial and ventricular septal defects can also be demonstrated by 2-D echocardiography and shunting across these defects can be demonstrated by Doppler echocardiography. Semilunar valves can be identified as pulmonary or aortic by following the great vessel until the bifurcation of the pulmonary artery or arch of the aorta is seen, this will help decide whether there is associated transposition of the great arteries. Measurement of peak Doppler flow velocities across the VSD and right ventricular outflow tract will not only reveal if obstruction is present at these sites but will also allow estimation of pulmonary artery pressures. Suprasternal notch imaging will be of use in demonstrating CoA, which is often seen in type II (transposition) patients. Contrast echocardiography with two-dimensional imaging will clearly demonstrate sequential opacification of the right atrium, left atrium, left ventricle and then the right ventricle. However, such a study is not always necessary for diagnosis.

6.3.4 Cardiac catheterization and selective cineangiography

The diagnosis of TA based on clinical, electrocardiographic, and echocardiographic features is relatively simple, and cardiac catheterization and selective cineangiography, rarely, if ever, are essential for arriving at the diagnosis. However, these procedures are useful and should be undertaken to resolve issues not clarified by non-invasive studies and to evaluate multiple physiologic and anatomic features prior to planned Fontan-Kreutzer operation (Rao 1992).

Oxygen saturation data reveal diminished systemic venous saturation; the extent of decrease is related to the systemic arterial desaturation and the severity of congestive heart failure. The pulmonary venous saturation is usually in the normal range. A significant decrease in left atrial saturation is expected because of obligatory right-to-left shunting across the patent foramen ovale. Falsely high or falsely low saturations may be measured in the left atrium because of streaming. The left ventricular saturations are usually well mixed and are more reliable. The saturations in the left atrium, left ventricle and aorta as well as those in the right ventricle and pulmonary artery are nearly equal. Systemic arterial (aortic) desaturation is always present and the extent of desaturation is proportional to the Qp:Qs.

The right atrial pressure may be mildly increased. If the foramen ovale is restrictive the pressure in the right atrium is markedly elevated; a mean pressure gradient of 5 mmHg across the patent foramen ovale in favor of the right atrium and giant "a" waves in the right atrium are indicative of an obstructive foramen ovale. The left atrial mean and left ventricular end-diastolic pressures are usually normal, but may be elevated if there is increased pulmonary blood flow, poor left ventricular function or significant mitral insufficiency. The right ventricular pressure is proportional to the size of the VSD in type I (normally related great arteries) patients while it is at systemic level in type II (transposition) patients. Systolic pressure gradient across the VSD may be seen if it is restrictive. The pulmonary artery pressure may be normal or increased depending upon the size of the VSD

(and associated PS) in type I patients and upon the presence or absence of subvalvar or valvar PS in type II patients. Aortic pressures are usually normal. If CoA is present, systolic hypertension and pressure gradient across the coarctation will be present.
Of all the calculated values, Qp:Qs and pulmonary vascular resistance are most useful. The Qp:Qs is diminished in type I hypoxemic patients with small or no VSD while it is markedly increased in type I patients with moderate to large VSDs and in most patients with type II anatomy. Pulmonary vascular resistance is an important factor to be taken into consideration for deciding to go ahead with Fontan-Kreutzer operation; elevated resistance adversely affects the outcome of the operation.
Selective right atrial angiography will confirm the diagnosis. Following right atrial injection, successive opacification of the left atrium and left ventricle without direct opacification of the right ventricle occurs and this negative shadow of the unopacified right ventricle, the so called right ventricular window is considered characteristic for TA. Selective left ventricular angiography is also recommended and is useful in evaluating its size and function, size and type of VSD, anatomy and size of the right ventricle, relationship of the great arteries and the source of pulmonary blood flow. Selective right ventricular and pulmonary arterial angiograms are possible with the currently available catheter/guide wire technology and may be necessary in some cases for better definition prior to considering “corrective” surgical procedures.

6.3.5 Management

Physiologically "corrective" operation for TA, namely, Fontan-Kreutzer procedure and its modifications, have improved the prognosis of patients with tricuspid atresia. Such physiologic correction is usually performed in patients older than 2 years. As stated above, most patients with TA present with symptoms in the neonatal period and should be effectively palliated to enable them to reach the age at which surgical correction could be undertaken. The objective of any management plan, apart from providing symptomatic relief and increased survival rate, should be to preserve, protect, and restore anatomy (good sized and undistorted pulmonary arteries) and physiology (normal pulmonary artery pressures and preserved left ventricular function) to normal such that a "corrective" procedure could later be performed safely.
Medical management of the neonate, just as for TGA patients, includes maintenance of neutral thermal environment, normal acid-base status, normoglycemia and normocalcemia by appropriate monitoring and correction, if needed. No more than 0.4 FI02 is necessary unless there is associated pulmonary pathology.
In neonates with low arterial PO_2 and O_2 saturation with ductal dependent pulmonary blood flow, the ductus should be kept open by intravenous infusion of PGE_1, in doses similar to that described in TOF and TGA sections. Once the infant is stabilized and appropriate diagnostic studies are performed, a Blalock-Taussig type of shunt is performed in the group with pulmonary oligemia.
In neonates and infants with pulmonary plethora and congestive heart failure, aggressive anticongestive therapy must be instituted. In type I (normally related great arteries) patients, the natural history of the VSD is such that it closes spontaneously and the infants will go on to develop pulmonary origemia (Rao 1977b). Because of these reasons, it is recommended that banding of the pulmonary artery not be routinely performed in this group of patients. If optimal anticongestive therapy with some delay does not produce adequate relief of

symptoms, pulmonary artery banding should then be considered. Alternatively, an absorbable band may be used (Rao 2001). By contrast, in type II (transposition) patients, banding of the pulmonary artery should be performed once the infant is stabilized with anticongestive measures. If there is associated CoA, it should also be relieved.
In infants with evidence for interatrial obstruction, balloon and/or blade atrial septostomy may be necessary.
Following initial palliation, the children should be followed under close cardiologic supervision. Currently, preferred "corrective" procedure is staged total cavopulmonary anastomosis. A bi-directional Glenn procedure (superior vena cava to pulmonary artery anastomois) may be performed around the age of six months. Preoperative catheter evaluation to define the pulmonary artery pressure and anatomy and to exclude a persistent left superior vena cava (because it may divert blood away from the pulmonary arteries) prior to bidirectional Glenn surgery should be undertaken. At the time of bidirectional Glenn procedure, stenoses, if any, of the pulmonary artery should be repaired. Issues related to subaortic obstruction and mitral valve regurgitation should also be addressed.
When the patients reach the age and size (approximately 15 Kg) suitable for Fontan-Kreutzer operation, diversion of inferior vena caval blood into the pulmonary artery either by a lateral tunnel or extracardiac conduit is recommended. At the present time extracardiac conduit diversion of inferior vena caval blood into the pulmonary artery) is preferred by most surgeons.. Immediately prior to Fontan conversion, cardiac catheterization should be undertaken to ensure normal anatomy and pressure of the pulmonary artery as well as normal left ventricular end-diastolic pressure. At the same time, aortopulmonary collaterals should be evaluated by means of selective subclavian artery and descending thoracic aortic angiography. If collateral vessels are present, they should be occluded with coils or devices, as appropriate.
In patients with transposition of the great arteries, early pulmonary artery banding, treatment of aortic coarctation, and relieving or bypassing subaortic obstruction should also be incorporated into the treatment plan.
If the patient has risk factors for poor outcome (for e.g., elevated pulmonary pressure/resistance, pulmonary artery distortion, and left ventricular dysfunction) for the corrective procedure, a fenestrated Fontan procedure should be considered. Some surgeons prefer fenestration for all patients. Six to twelve months later, transcatheter closure of the fenestration may be undertaken if the fenestration did not spontaneously close.
Close follow-up after correction is indicated. While most of these patients will do well, some may develop arrhythmia (atrial flutter or fibrillation, paroxysmal supraventricular tachycardia), obstructed Fontan pathways, branch pulmonary artery stenosis, thrombo-embolism, persistent right to left shunts (Fontan fenestrations or atrial septal defects), systemic venous to pulmonary venous collateral vessels and protein-losing enteropathy. Detailed evaluation of these problems and appropriate treatment is mandatory

7. Conclusions

Congenital heart defect is an anatomic malformation of the heart and/or great vessel, which occurs during intrauterine development. The incidence of CHD is 0.6 to 0.8% of live-births. The exact etiology of CHD is not known and the majority of cardiac defects can be explained by multifactorial inheritance hypothesis. The CHD may be classified as acyanotic and cyanotic defects and the former is further divided into obstructive and left-to-right shunt

lesions. Pathologic, physiologic, clinical and laboratory features of nine most common CHD, described in this chapter are distinctive. Methods of management for each of these defects are outlined. Based on this review, it appears that while the etiology of CHD is not clearly identified, their recognition by clinical evaluation and non-invasive laboratory tests is possible and their treatment with currently available transcatheter and surgical methods is feasible, effective and safe.

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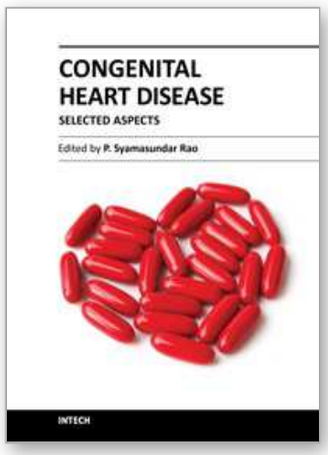

Congenital Heart Disease - Selected Aspects

Edited by Prof. P. Syamasundar Rao

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There are significant advances in the understanding of the molecular mechanisms of cardiac development and the etiology of congenital heart disease (CHD). However, these have not yet evolved to such a degree so as to be useful in preventing CHD at this time. Developments such as early detection of the neonates with serious heart disease and their rapid transport to tertiary care centers, availability of highly sensitive noninvasive diagnostic tools, advances in neonatal care and anesthesia, progress in transcatheter interventional procedures and extension of complicated surgical procedures to the neonate and infant have advanced to such a degree that almost all congenital cardiac defects can be diagnosed and "corrected". Treatment of the majority of acyanotic and simpler cyanotic heart defects with currently available transcatheter and surgical techniques is feasible, effective and safe. The application of staged total cavo-pulmonary connection (Fontan) has markedly improved the long-term outlook of children who have one functioning ventricle. This book, I hope, will serve as a rich source of information to the physician caring for infants, children and adults with CHD which may help them provide optimal care for their patients.

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