Symposium on Advances in Cardiology - I

Diagnosis and Management of Cyanotic Congenital Heart Disease: Part I

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ABSTRACT

Most commonly encountered cyanotic cardiac lesions in children, namely, tetralogy of Fallot, transposition of the great arteries and tricuspid atresia are reviewed. Pathology, pathophysiology, clinical features, non-invasive and invasive laboratory studies and management are discussed. The clinical and non-invasive laboratory features are sufficiently characteristic for making the diagnosis and invasive cardiac catheterization and angiographic studies are not routinely required and are needed either to define features, not clearly defined by non-invasive studies or as a part of catheter-based intervention. Surgical correction or effective palliation can be undertaken with relatively low risk. However, residual defects, some requiring repeat catheter or surgical intervention, are present in a significant percentage of patients and therefore, continued follow-up after surgery is mandatory. **[Indian J Pediatr 2009; 76(1) : 57-70]** *E-mail: P.Syamasundar.Rao@uth.tmc.edu*

Key words : *Cyanotic cardiac lesions; Diagnosis; Invasive cardiac catheterization; Surgical intervention*

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. Congenital heart defects may be classified into acyanotic and cyanotic depending upon whether the patients clinically exhibit cyanosis. The acyanotic defects (obstructive and left-to-right shunt lesions) were discussed in a previous Pediatric Cardiology Symposium in this Journal[1,2] and will not be reviewed here. 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 CHD 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 1. This review consists of two parts; in the first part the objective is to describe the important findings in history, physical examination and laboratory studies of the most common cyanotic lesions, namely tetralogy of Fallot, transposition of the great arteries and tricuspid atresia and to discuss the available options in the management of these defects. In the second part a brief discussion of the other cyanotic CHD will be included.

Table 1. Common Cyanotic Congenital Heart Defects (5 Ts)

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

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.[3] Fallot defined it as a constellation of four abnormalities to include a ventricular septal defect (VSD), pulmonary stenosis (PS), right ventricular hypertrophy and dextroposition of the aorta. The ventricular defect is always large and

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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 and/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 intramyocardial.

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 resultant cyanosis and hypoxemia stimulate bone marrow (*via* kidney and erythropoietin) and produce polycythemia.[4] While the polycythemia is helpful in increasing oxygen carrying capacity, it becomes counter-productive when the hematocrit is excessive (> 60 to 70%).

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 on. 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 catecholamines 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.[5] 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

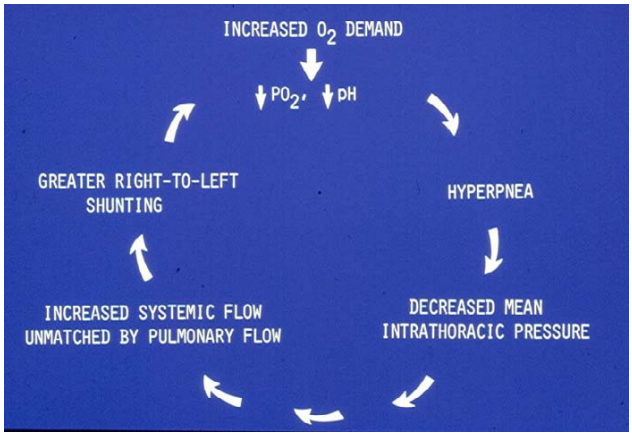

Fig. 1. The mechanism of hypercyanotic spells as proposed by Guntheroth.[5] See the txt for details.

consumption before the above described 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 (Fig. 1). 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.[4,5] Most workers believe that this is the most likely mechanism for the development of spells.

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, 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 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, usually heard on the back.

Noninvasive evaluation

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.

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

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.[4]

Echocardiogram is very helpful in confirming the diagnosis and in evaluating several of the issues related to TOF (Fig. 2). 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.

CT and MRI. These noninvasive studies, including

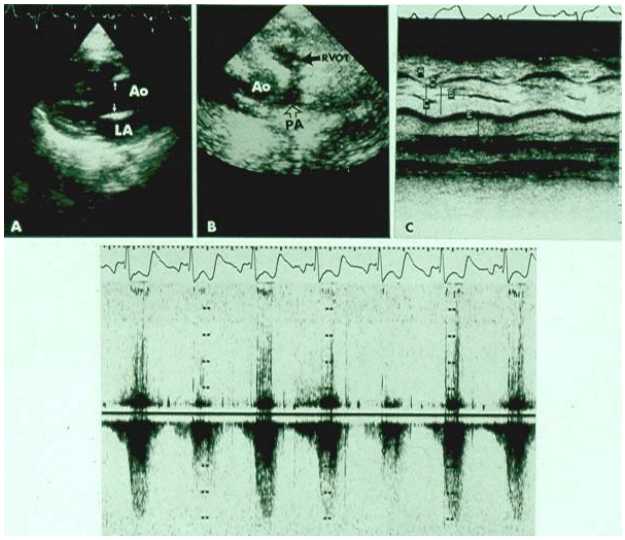

Fig. 2. Selected echocardiographic and Doppler recordings from a patient with tetralogy of Fallot, demonstrating a large aorta (Ao) overriding the interventricular septum (A), narrowed right ventricular outflow tract (RVOT) (B), large aorta and a small left atrium (LA) (C) and high Doppler flow velocity (D) across the RVOT. PA, pulmonary artery.

three-dimensional reconstruction, are useful in defining issues that could not be addressed by conventional echocardiography. But, the expertise in performing and interpreting such studies are not readily available in all institutions.

Cardiac catheterization and angiography: Catheterization is not routinely required, but may be performed if all the data required for making decision for surgical correction can not be obtained by non-invasive studies with reasonable certainty.

Oxygen saturation data reveal systemic venous and arterial desaturation, usually proportional to the degree of right ventricular outflow obstruction. There are usually no left-to-right shunts 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 non-restrictive 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.

Angiography is an integral part of cardiac catheterization. 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 (15^0 LAO and 35^0 cranial) 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 need be by selective coronary angiography. Some of the above angiographic features are illustrated in fig. 3.

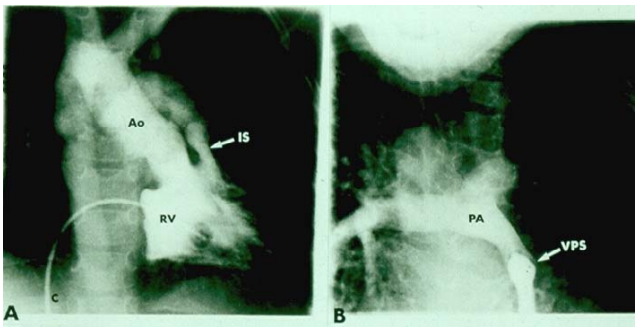

Fig. 3. A. Selected right ventricular (RV) cineangiographic frame demonstrating enlarged and trabeculated RV opacifying the aorta (Ao) through a ventricular septal defect (not shown) and severe infundibular stenosis (IS). Note that the AO descends on the right side of the spine. B. Selected RV outflow tract cineangiographic frame demonstrating markedly narrowed pulmonary valve (VPS) with doming. The main (PA) and branch pulmonary arteries are well visualized. C, catheter.

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.

Spells. Treatment of an infant with cyanotic spell may be summarized[5,6] as follows:

• 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.

• 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.

• 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.

• 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.

• Correction of metabolic acidosis (with sodium bicarbonate), anemia (by blood transfusion), and dehydration (by appropriate fluids), if present, is very important at this stage.

• 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. Alternatively, phenylephrine may be given to increase systemic vascular resistance.

• Another option, 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.

• Infrequently, general anesthesia may be necessary to break the spell.

• If the infant does not improve with any of the aforementioned measures, an emergency systemic-to-pulmonary artery shunt (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.[7-10] An alternative to surgery is oral propranolol (dosage as above) which may help postpone surgery by several months.

Surgery. 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[11,12] are options most surgeons prefer modified Blalock-Taussig shunt using an interposition Gore-Tex graft between right or left subclavian arteries to the ipsilateral pulmonary artery. We have used balloon pulmonary valvuloplasty[7-10] 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.[7-10]

Follow-up of all patients after surgery is important to address issues related to residual defects. On long-term follow-up many patients seem to require replacement of pulmonary valves for the management of pulmonary valve insufficiency.

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 neonatal 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), ventriculo-arterial discordance (aorta arising from the right ventricle and the pulmonary artery from the ventricle) and the aortic

valve is located to the right of pulmonary valve (d-TGA). The systemic venous blood from the vena cavae enters the right atrium and right ventricle and from there the aorta while the pulmonary venous blood enters the left atrium and left ventricle and from there the pulmonary artery (Fig. 4). 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 infants will not survive unless there are inter-circulatory shunts such as atrial or ventricular septal defect or patent ductus arteriosus.

Symptoms

Clinical features depend upon the anatomic type, namely Group I, TGA with intact ventricular septum;

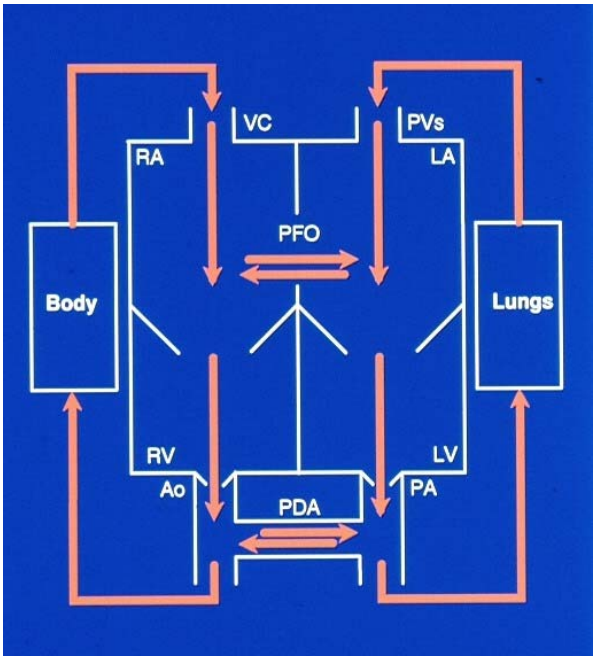

Fig. 4. Box diagram of the heart showing parallel circulations in transposition of the great arteries. Note that right ventricle pumps into the aorta (Ao) (because of transposition) which goes to the body and returns into right atrium and back into the body. Similarly left ventricular output goes to the pulmonary artery (PA) and lungs and returns back to the left atrium and left ventricle to be pumped back into the lungs. Unless there are inter-circulatory communications via either a patent foramen ovale or patent ductus arteriosus, the infant cannot survive. Mixing across a ventricular septal defect (VSD) if such is present (not shown in the diagram) would also prevent progressive hypoxemia and death.

Group II, TGA with VSD, and Group III, TGA with VSD and PS.

In group I with intact septum, the infants usually present with cyanosis within the first week of life (sometimes within hours to days of life). They may otherwise be asymptomatic. However, they will, with time, 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.

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/VI 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.

Noninvasive Evaluation

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 patients, 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.

The **electrocardiogram** 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.

Echocardiogram 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 (Fig. 5), 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 (Fig. 6). On-end visualization of both the aorta and pulmonary artery simultaneously on a precordial short axis view of the heart is also helpful in suggesting TGA. The presence of an interatrial 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.

Blood gas values are useful in demonstrating the degree of hypoxemia and ventilatory status.

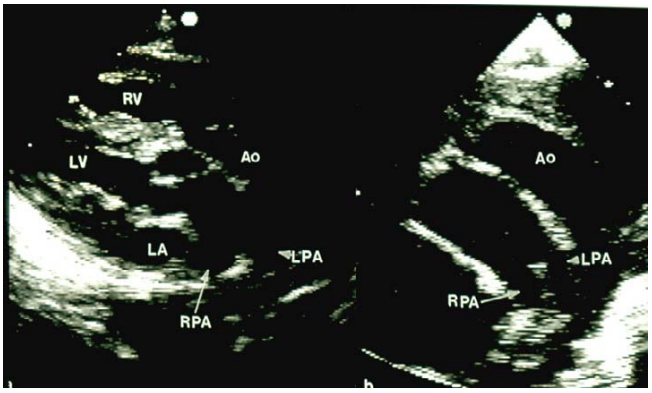

Fig. 5. Selected video frames from a 2-dimensional echocardiographic views of an infant with transposition of the great arteries. In a, note the great vessel coming off of the left ventricle (LV) courses posteriorly and bifurcates into left (LPA) and right (RPA) pulmonary arteries. In b. posterior vessel is similarly seen to bifurcate. The anterior vessel is aorta (Ao). LA, left atrium; RV, right ventricle.

Hemoglobin and hematocrit are particularly useful in the follow-up of older children.

Cardiac catheterization and angiography

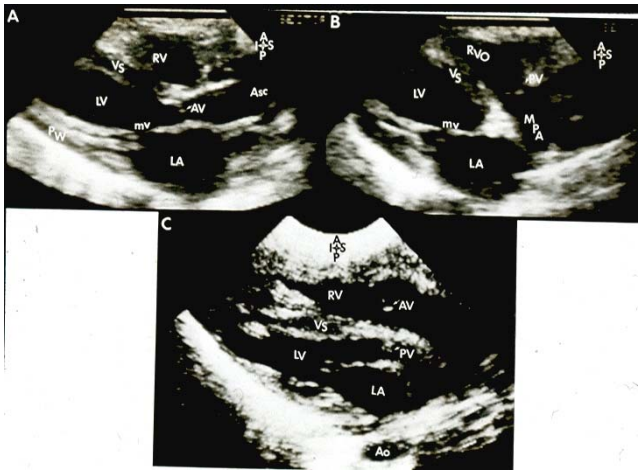

Fig. 6. Precordial long axis views of two neonates, first (top, A & B) with normally related great arteries and the second (bottom, C) with transposition of the great arteries. A. Note that the posterior vessel arising from the left ventricle (LV) courses somewhat anteriorly, indicating that it is likely to be the aorta. In B, the anterior vessel coming of off the right ventricle (RV) divides into right and left pulmonary arteries, suggesting that his vessel is main pulmonary artery (MPA). In C, the posterior vessel is coursing backward (posteriorly) after its origin from the LV and is likely to be the MPA, suggesting transposition of the great arteries. Ao, aorta; Asc, ascending aorta; AV, aortic valve; LA, left atrium; mv, mitral valve; PV, pulmonary valve; PW, posterior wall of LV; RVO, right ventricular outflow tract; VS, ventricular septum.

With the increased 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 pressures 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 (Fig. 7) 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 coarctation of the aorta. Left ventricular cineangiogram (Fig. 7) 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.

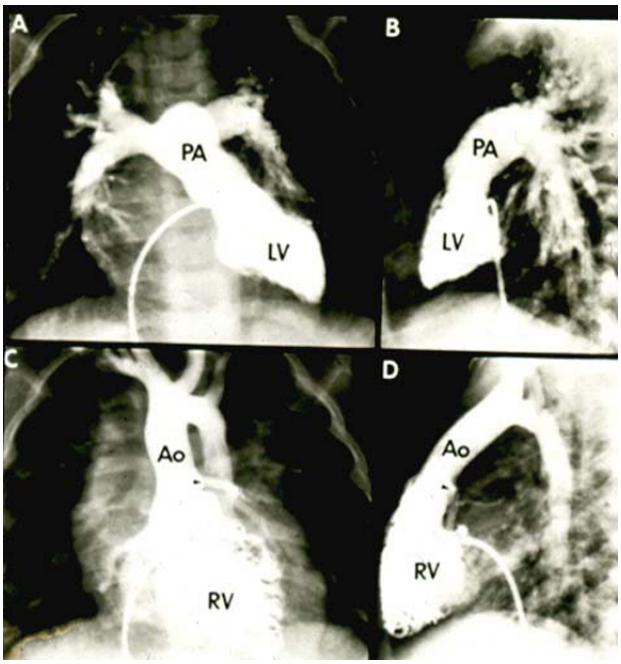

Fig. 7. A & B. Left ventricular (LV) cineangiogram in postero-anterior (A) and lateral (B) views demonstrating a finely trabeculated, morphologic left ventricle with prompt opacification of the pulmonary artery (PA). The PA is inferior and posterior to its usual position. C & D. Right ventricular (RV) cineangiogram in postero-anterior (C) and lateral (D) views showing a coarsely trabeculated ventricle with opacification of the aorta (Ao). Note that the aortic valve is superior and anterior (D) to its usual position.

Management

The initial management of TGA 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.

Untreated, TGA with intact septum carries a poor prognosis. If an infant is getting progressively hypoxemic, it is likely that the inter-circulatory pathways (patent foramen ovale and patent ductus arteriosus) are closing. Prostaglandin E_1 (PGE_1) (0.05 to 0.1 mcg/kg/min) intravenously may help open the ductus, thus improve oxygenation. Balloon atrial septostomy[13,14] (Fig. 8) may be necessary to improve hypoxemia even after PGE_1. Total surgical correction by arterial switch procedure (Jatene)[15] is the treatment of choice in these neonates and will be discussed here-

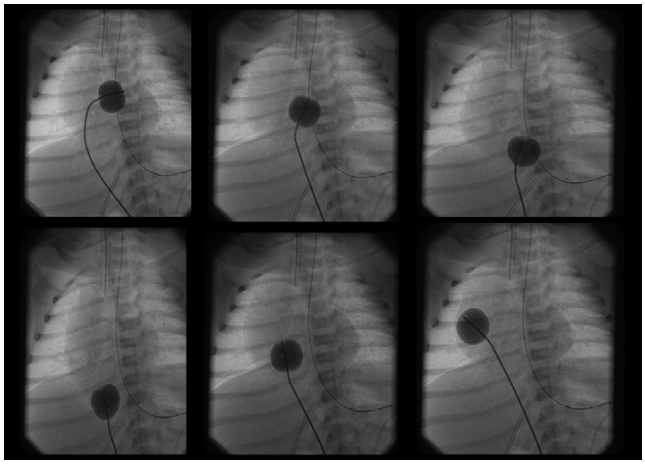

Fig. 8. Selected cinfluroscopic frames of the Rashkind's balloon septostomy procedure. Note the position of the inflated balloon in the left atrium (A) and in right atrium and inferior vena cava in successive frames, as it is rapidly and forcefully withdrawn across the atrial septum (B,C, & D). After it reaches the inferior vena cava (D), it is rapidly advanced into the right atrium (E & F) in order not to inadvertently occlude the inferior vena cava in case of failure to deflate the balloon (which is quite rare).

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 types of 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.

Surgical correction. 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 intraatrial 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 the 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. Again, follow-up after surgery is mandatory to manage residual defects.

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. Most issues related to this disease entity were extensively reviewed elsewhere;[16-20] the interested reader is referred to these publication for additional details. 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 interatrial 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, transposition of the great arteries other than d-transposition; and Type IV, Truncus arteriosus (Table 2).[21] 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

TABLE 2. Unified Classification of Tricuspid Atresia

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
	Subtype 1. L-transposition of the great arteries	
	Subtype 2. Double outlet right ventricle	Subgroup c. Normal pulmonary arteries (no pulmonary stenosis)
	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		

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.

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.[17,19,20] 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 is well known[22] and coarctation, when present, makes them vulnerable to early cardiac failure.

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 interatrial 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 coarctation of the aorta), minimal cyanosis and hepatomegaly. Prominent "a" waves in the jugular veins and/or presystolic hepatic pulsations may be observed with associated interatrial 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.

Non-invasive evaluation

Chest X-ray 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.

The **electrocardiogram** (Fig. 9) 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.

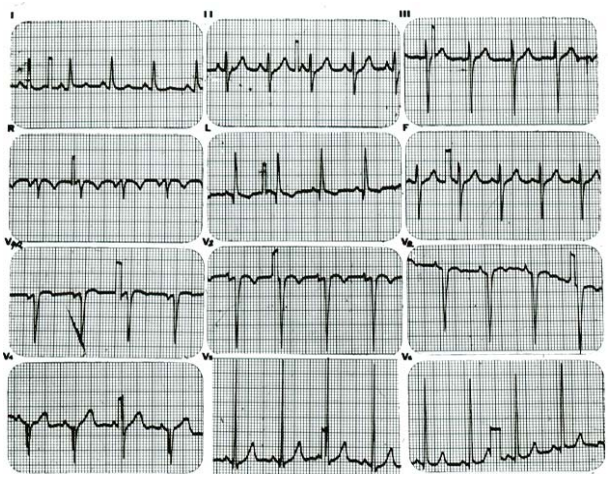

Fig. 9. A 12-lead electrocardiogram of a child with tricuspid atresia, demonstrating a mean frontal plane vector (axis) of -45^0. Also, note tall R waves in the left chest leads and deep S waves in the right chest leads indicative of left ventricular hypertrophy and tiny, if any r waves in right chest leads, suggestive of small right ventricle, all typical features tricuspid atresia.

Echocardiogram 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 (Fig. 10). 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 interatrial 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

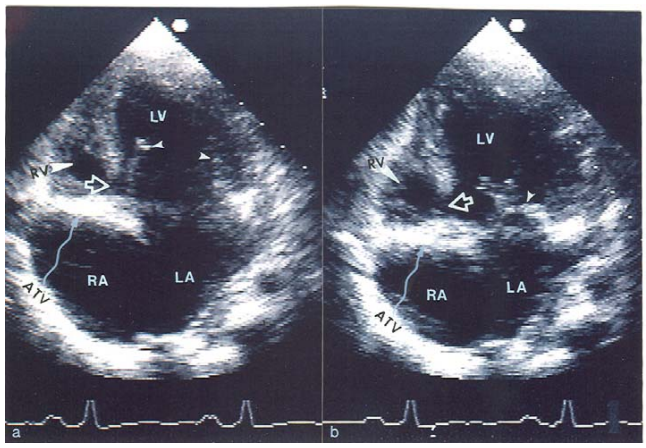

Fig. 10. Apical four chamber two-dimensional echocardiographic views of a child with tricuspid atresia showing an enlarged left ventricle (LV), a small right ventricle (RV) and a dense band of echoes, representing atretic tricuspid valve (ATV) at the site where the tricuspid valve echo should be. Atrial (not labeled) and ventricular (open arrow-head) septal defects are also seen. Note the attachment of the detectable atrioventricular valve to the left side of the interatrial septum. Frames a and b show open and closed mitral valve leaflets (closed arrow-heads) respectively. LA, left atrium; RA, right atrium.

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 obstruction if present at these sites but will also allow estimation of pulmonary artery pressures. Suprasternal notch imaging will be of use in demonstrating aortic coarctation, 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.

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.[23]

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 largely proportional to the Qp:Qs (has a curvilinear relationship[23]).

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 aortic coarctation is present, systolic hypertension and pressure gradient across the coarctation may 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 VSD 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 not only confirm the diagnosis but also gives information regarding its size and location and shape of the atrial appendage. Following right atrial injection, successive opacification of the left atrium and left ventricle (Fig. 11) occurs without direct opacification of the right ventricle 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

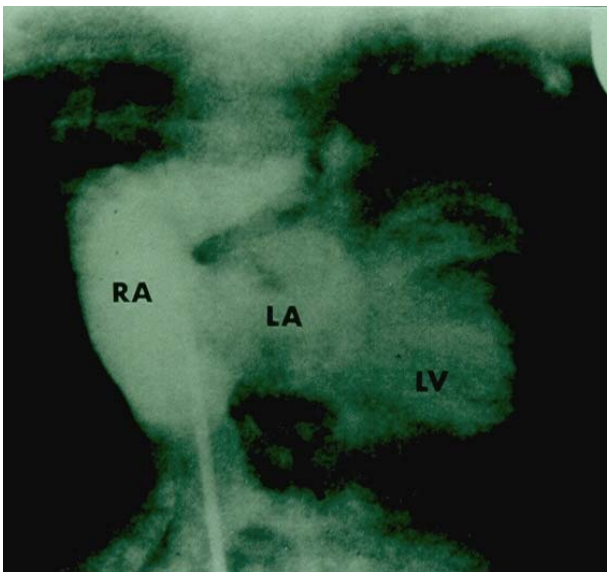

Fig. 11. Selected frame from a postero-anterior view of a right atrial (RA) angiogram in an infant with tricuspid atresia showing successive opacification of the left atrium (LA) and left ventricle (LV). There was no direct and immediate opacification of the right ventricle; the negative shadow, the so called right ventricular window, is shown with an arrow.

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.

Management

Physiologically "corrective" operation for TA, namely, Fontan-Kreutzer procedure[24,25] 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 $FI0_2$ 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[26,27] and the infants will go on to develop pulmonary origemia. Because of these reasons, it is recommended that banding of the pulmonary artery not be routinely performed in this group of patients. If optimal anti-congestive therapy with some delay does not produce adequate relief of symptoms, pulmonary artery banding should then be considered. Perhaps a serious consideration for using absorbable band material[28-29] should be given. An absorbable pulmonary artery band has been used for palliation in such infants.[28] By restricting the pulmonary blood flow, the absorbable polydioxanone band decreases pulmonary artery pressure initially and helps abate symptoms of heart failure. As the VSD spontaneously closes, the pulmonary artery band gets resorbed and does not produce the severe pulmonary oligemia[29] that might have been associated with a conventional non-absorbable band. This is an inventive approach, although it is likely to be helpful in a limited number of patients.[29] By contrast, in type II (transposition) patients, banding of the pulmonary artery should be performed once the infant is stabilized with anti-congestive measures. If there is associated aortic coarctation, 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 procedure is staged total cavo-pulmonary anastomosis.[30] Between the ages of 6 months to 1 year, bidirectional Glenn (the superior vena cava is divided and anastomosed to the right pulmonary artery, end to side so that the blood form the superior vena cava flows into both the right and left pulmonary arteries) is performed. The bidirectional Glenn shunt may be performed in patients as young as 3 months; however, an increased probability of failure exists at this young age, presumably related to pulmonary vascular reactivity and we do not advocate such an approach. At the time of bidirectional Glenn, repair of pulmonary artery narrowing, if present, should be undertaken. If the atrial septal defect is restrictive it should be enlarged. Issues related to subaortic obstruction and mitral valve regurgitation should also be addressed. At the age of 2 years or when the patient reaches a weight of 15 kg, Fontan conversion is performed. Two types of Fontan conversion are described, namely intra-atrial tunnel diverting the inferior vena caval flow into the pulmonary arteries and an extracardiac conduit, again diverting the inferior vena caval flow into the pulmonary arteries. Extracardiac Fontan is preferred by most surgeons. In patients with associated transposition of the great arteries, early pulmonary artery banding, relief of aortic coarctation (if present), and bypassing (by Damus-Kaye-Stansel) or resecting subaortic obstruction should be incorporated into the management plan.

Before Fontan conversion, cardiac catheterization should be undertaken to ensure normal pulmonary artery anatomy and pressure 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 coil occluded. 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. Transcatheter closure of the fenestration may be undertaken at a later date. Two-stage staged surgical-catheter approach has been advocated;[31] initially a modified hemi-Fontan is performed that is later completed by transcatheter methodology. This reduces the total number of operations required. However, there is only a limited experience with this approach.

Periodic follow-up after surgical correction is necessary for continued medical management, but also to address complications that are not uncommon after Fontan surgery.[20]

CONCLUSION

In this review, tetralogy of Fallot, transposition of the great arteries and tricuspid atresia, the three most common cyanotic congenital heart defects in children, are discussed. The clinical presentation, laboratory findings and management options are described. These defects have sufficiently distinctive features such that they can be diagnosed with relative ease. Upon diagnosis, some require immediate treatment for stabilization and all require subsequent corrective or palliative surgical therapy. Significant post-operative residual abnormalities may be present and some may require catheter interventional procedures and/or repeat surgery. Consequently periodic follow-up is mandatory.

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