NEONATOLOGY TODAY

News and Information for BC/BE Neonatologists and Perinatologists

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IN THIS ISSUE

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Annual International Neonatal Conference
Jun. 14-16, 2012;
Billingham, Teesside Valley, UK
www.neonatalconference.co.uk/home.html

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Annual International Neonatal Conference
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Corporate Offices:
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Mailing Address:
PO Box 444
Manzanita, OR 97131 USA

Editorial and Subscription Offices
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Neonatology Today (NT) is a monthly newsletter for Neonatologists and Perinatologists that provides timely news and information regarding the care of newborns and the diagnosis and treatment of premature and/or sick infants.

Tricuspid Atresia in the Neonate

By P. Syamasundar Rao, MD and Srilatha Alapati, MD

Introduction

In the previous issues of *Neonatology Today*, we discussed general topics of congenital heart disease in the neonate,[1-5] but began addressing individual cardiac lesions[6-8] recently. In this issue, tricuspid atresia will be discussed.

Tricuspid Atresia

Tricuspid atresia is a cyanotic, congenital cardiac anomaly and is defined as congenital absence or agenesis of the morphologic tricuspid valve.[9,10] It is the third most common cyanotic congenital heart defect and is the most common cause of cyanosis with left ventricular hypertrophy. Whereas there is a controversy with regard to terminology (tricuspid atresia, univentricular heart or univentricular connection), the authors are of the opinion that the term "tricuspid atresia" is the correct and logical term to describe this well-characterized pathologic and clinical entity; the reasons are detailed elsewhere.[10-12]

A thorough review by Rashkind[13] indicates that the first documented case of tricuspid atresia was that of Kreysig in 1817,[14] although the 1812 report by the editors of *London Medical Review*[15] appears to fit the description of tricuspid atresia.

The true prevalence of tricuspid atresia is not known. Extensive review of the literature revealed an autopsy prevalence rate of 2.9% and a clinical prevalence rate of 1.4% among subjects with congenital heart disease.[16] The clinical prevalence of tricuspid atresia in neonates with congenital heart defects is also similar at 1.5%.[17] With the known prevalence of congenital heart defects of 0.8% of live births, it is estimated that tricuspid atresia occurs approximately 1 in 10,000 live births.[16] There is not a gender preponderance for tricuspid atresia, but male preponderance appears to be present in tricuspid atresia patients with associated transposition of the great arteries: male to female ratio was 2:1.[16,18]

In this paper, we will discuss: classification, anatomic, physiologic and clinical features, noninvasive evaluation, differential diagnosis, management and prognosis of tricuspid atresia in the neonate.

Classification

Tricuspid atresia may be classified on the basis of valve morphology,[19] appearance of pulmonary

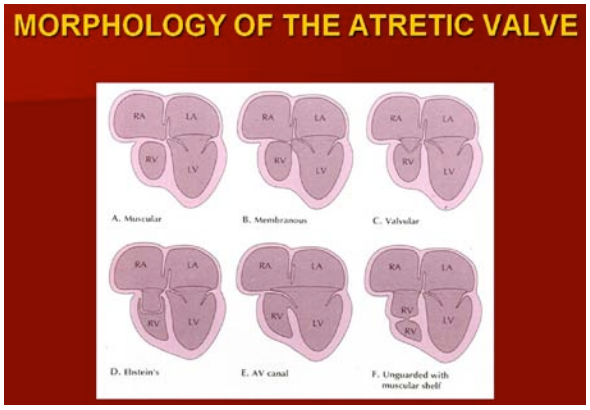

Figure 1. Diagrammatic portrayal of anatomic types of tricuspid atresia based on the morphology of the atretic tricuspid valve. a. muscular type, b. membranous type, c. valvular type, d. Ebstein's type, e. atrioventricular canal type, and f. unguarded valve with muscular shelf. For the sake of simplicity, great vessels are not shown. Also note that no ventricular septal defects are shown. LA, left atrium; LV, left ventricle; RA, right atrium; RV right ventricle.

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vascular markings on chest X-ray,[20] and associated cardiac defects.[9,21-24]

Looking at the morphology of the atretic tricuspid valve (Figure 1), it may be classified into muscular, membranous, valvular, Ebstein's, unguarded with muscular shelf and atrioventricular canal types.[19,24-26] The muscular type, constituting 89% of cases,[24,26] is the most common type; the remaining types account for 11% of cases.

A classification based solely on the x-ray appearance of pulmonary vascular markings was put forward by Astley:[20] Group A. Decreased pulmonary vascular markings, and Group B. Increased pulmonary vascular markings. Dick and his associates[18] added another group to Astley's classification: Group C. Transition from increased to decreased pulmonary vascular markings in serial chest films. The above three classifications have clinical value but a classification based on associated cardiac defects appears to be more useful clinically.[9,24]

A classification based on great artery interrelationship was first proposed by Kühne in 1906.[21] The classification was later refined by Edwards and Burchell[22] and by Keith, Rowe, and Vlad.[23,27,28] Because of some apparent inconsistencies in sub grouping and the need for inclusion of all variations in great artery anatomy, we proposed a new, unified classification,[9,29] and this is listed in Table I. First, the tricuspid atresia is classified into four major types based on the great artery relationship. Each type is identified with a Roman numeral: 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. The type III is again subdivided into several subtypes (see Table I) and is identified with an Arabic number (1 thru 5). Each type and subtype are further divided into subgroups on the basis of pulmonary arteries; each subgroup is indicated by a lower case letter: Subgroup a – Pulmonary atresia, b – Pulmonary stenosis or hypoplasia, and c – Normal pulmonary arteries (no pulmonary stenosis).[9,24,26,29] Once a patient with tricuspid atresia is thus classified, the status of the interventricular septum, i.e., intact, small or large ventricular septal defect (VSD), or multiple VSD's and other associated malformations should be described. If one wants to follow the terminology of congenital heart disease proposed and re-emphasized by Van Praagh,[30] one could include the remaining segmental subsets, namely visceroatrial situs and ventricular loop. Each case could be described by notations {S,D,S}, {S,D,D}, {S,D,L} and so on as the case may be.[30,31]

Table 1.
A 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 • 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)	Subgroup c. Normal pulmonary arteries (no pulmonary stenosis)
Type IV	Persistent truncus arteriosus	

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Pathologic Anatomy

The most common type of tricuspid atresia, muscular variety, is characterized by a dimple or a localized fibrous thickening in the floor of the right atrium (Figure 2) at the expected site of the tricuspid valve[23] and constitutes 89% of the cases.[24,26] No valvar material can be identified either by gross or microscopic examination.[23] Other anatomic types, namely, membranous type (6.6%) with the atrioventricular portion of the membranous septum forming the floor of the right atrium,[32,33] valvular type (1%) with minute valve cusps which are fused,[23,34,35] Ebstein's type (2.6%) with Ebstein's deformity of the tricuspid valve leaflets with fusion of the valve leaflets,[18,32,36] common atrioventricular canal type (0.2%) in which a leaflet of the common atrioventricular canal completely seals off the only entry into the right ventricle,[25,37] and unguarded type (0.6%) with muscular shelf[38] have also been described and are diagrammatically portrayed in Figure 1. For further details of valve morphology types of tricuspid atresia the reader is referred to our previous reviews.[24,26]

With tricuspid atresia, the right atrium is usually enlarged and its wall thickened and hypertrophied. The interatrial communication, which is necessary for survival, is usually a stretched patent foramen ovale; sometimes an ostium secundum atrial septal defect and rarely an ostium primum atrial septal defect may be present. Occasionally the interatrial communication is obstructive and may form an aneurysm of the fossa ovalis causing obstruction to the mitral flow. 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

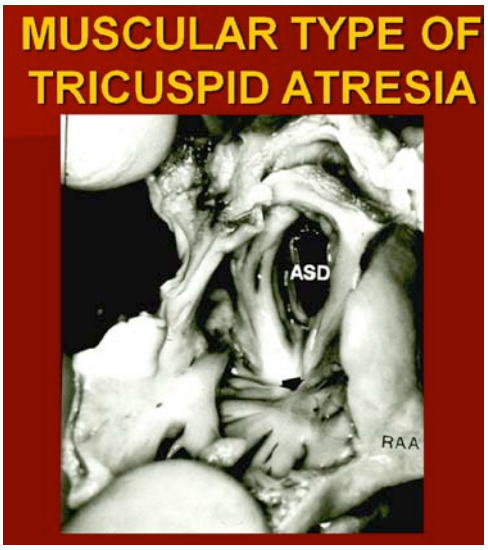

Figure 2. Heart Specimen of a patient with muscular type of tricuspid atresia; the right atrium is opened by cutting through the right atrial appendage (RAA). Note dimple (arrow) in the floor of the right atrium with muscle fibers radiating around it. An atrial septal defect (ASD) is also shown. The impression that one gets from the literature is that this dimple is present in most cases of tricuspid atresia. Careful inspection of the heart specimen by several investigators suggests that this dimple is seen in only 29 to 83% of muscular type of tricuspid atresia cases.

is clearly a morphologic left ventricle with only occasional abnormalities;[32] however, it is enlarged and hypertrophied.

The VSD may be large, small or non-existent (intact ventricular septum), or multiple VSDs may be present. When present, it may be:

a) conoventricular or perimembranous (located inferior to the septal band),
b) conal septal malalignment VSD (located in between the anterosuperior and posteroinferior limbs of septal band),
c) muscular (located inferiorly when compared to a and b), and
d) atrioventricular canal type.[31]

In the author's experience, muscular VSDs are most common.[39-42]

Also, most of these VSDs are restrictive and produce sub-pulmonary stenosis in the Type I patients and subaortic stenosis in the Type II patients.[39-47]

The right ventricle is small and hypoplastic; even the largest of the right ventricles that are present in patients with large VSDs and/or transposition of the great arteries are smaller than normal. It may be extremely small so that it may escape detection on gross examination of the specimen as in Type Ia cases. It can be identified at the right upper aspect of the ventricular mass. On occasion, it can be identified

only on microscopic examination.[22,23] However, in most cases the ventricle is a true right ventricle[32,48] consisting of:

a) a sharply demarcated infundibulum with septal and parietal bands and
b) a sinus with trabeculae which communicates with the left ventricle via a VSD.

The inflow region of the right ventricle, by definition, is absent; although papillary muscles may be present occasionally.[28]

The relative position of the great vessels is quite variable and has been the basis for classification of this anomaly, which has been described above. The ascending aorta may be normal in size or large. Pulmonary outflow obstruction may be either subvalvar or valvar in patients with transposition of the great arteries, while in patients with normally related great arteries the pulmonary obstruction is often at the VSD level In a few cases, subvalvar pulmonary stenosis, narrow tract of the hypoplastic right ventricle and, rarely, valvar pulmonary stenosis may also be responsible for pulmonary outflow tract obstruction. With pulmonary atresia, either a patent ductus arteriosus or aortopulmonary collateral vessels may be present.

A large number of additional abnormalities may be present in 30% of tricuspid atresia patients.[49,50] Significant among these are persistent left superior vena cava and coarctation of the aorta; the latter is much more common in Type II (transposition) patients. The possible physiologic reason for the latter is discussed in the next section.

Pathophysiology

Prenatal Circulation

Tricuspid atresia is not detrimental to normal fetal development. In a normally formed fetus, the highly saturated inferior vena caval blood is preferentially shunted into the left atrium via the patent foramen ovale, and from there into the left ventricle and aorta. The superior vena caval blood containing desaturated blood is directed towards the tricuspid valve and right ventricle, and from there into the pulmonary arteries, ductus arteriosus, and descending aorta. Thus, in a normal fetus, the head, heart and upper extremities are supplied with blood at higher PO_2 while the lungs, the lower part of the body, and placenta are perfused by blood with lower PO_2. In tricuspid atresia, both vena caval streams have to be shunted across the foramen ovale into the left atrium and left ventricle. Therefore, the PO_2 differential to various parts of the body that is normally present does not exist. Whether this higher PO_2 to the lungs influences the pulmonary arteriolar smooth muscle development or not, is not known.[51] The lower than normal PO_2 to the brain and upper part of the body does not seem to impair their development, at least as observed clinically.

In Type I (normally related great arteries) patients with intact ventricular septum and/or pulmonary atresia (Type Ia) and Type II (transposition of the great arteries) patients with pulmonary atresia (Type IIa), the pulmonary blood flow must be derived entirely through the ductus. Since the ductus is carrying only the pulmonary blood flow, representing 8 to 10% of the combined ventricular output in contradistinction to 66% in the normal fetus,[1,51] the ductus arteriosus is likely to be smaller than normal. This and the acute angulation of the ductus at its aortic origin because of reversal of direction of ductal flow may render the ductus less responsive to the usual post-natal stimuli.[51]

In Type I patients with VSD, the amount of anterograde blood flow from the left ventricle through the VSD into the right ventricle, the pulmonary artery, and ductus arteriosus versus the amount of blood flow retrograde from the aorta to the ductus arteriosus varies with size of the VSD. The larger the VSD, the greater is the quantity of anterograde ductal flow.

In Type I patients with a small or no VSD, most of the left ventricular blood is ejected into the aorta which is then carried to the entire body including the placenta and lower part of the body. Thus, the aortic isthmus carries a larger proportion of ventricular output than normal; presumably, this is the reason for the rarity of aortic coarctation in these subgroups of tricuspid atresia patients. In Type II (transposition) patients without significant pulmonary stenosis, because the VSD is usually smaller than the pulmonary valve ring,[52] a larger proportion of blood traverses the pulmonary artery and ductus arteriosus, and therefore, the aortic isthmus flow decreases, thus accounting for higher incidence of aortic coarctation and aortic arch anomalies seen with these types of tricuspid atresia.

Postnatal Circulation

An obligatory right-to-left shunt occurs at the atrial level in all types and subtypes of tricuspid atresia (Figure 3). Usually, this shunting is through a patent foramen ovale, but on occasion, secundum or primum atrial septal defects may be present. Thus, the systemic and coronary venous blood mixes with pulmonary venous return in the left atrium. These mixed pulmonary, coronary and systemic venous returns enter the left ventricle.

In Type I (normally-related great arteries) patients with a VSD, left-to-right ventricular shunt occurs, thus perfusing the lungs (Figure 4, left panel). In the absence of a VSD, the pulmonary circulation is derived either via a patent ductus arteriosus (Figure 4, right panel) or through broncho-pulmonary or persistent embryonic aortopulmonary collateral vessels. The presence of either a VSD or other means of blood supply to the lungs is crucial for the patient's survival. The aortic blood flow is derived directly from the left ventricle.

In Type II (D-transposition of the great arteries) patients, the pulmonary blood flow is directly derived from the left ventricle. The systemic blood flow is via the VSD and the right ventricle. In Type III, Subtype 1 with L-transposition

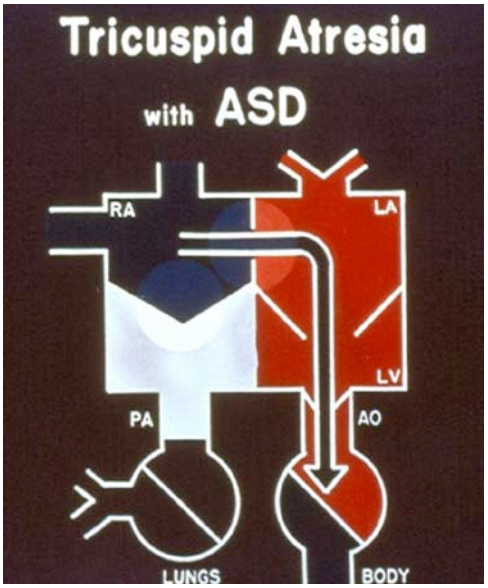

Figure 3. Box diagram of tricuspid atresia. The systemic venous return cannot exit into the right ventricle and its egress has to be into the left atrium (LA) via a patent foramen ovale or an atrial septal defect (ASD). Then, the blood traverses into the left ventricle (LV) and aorta (Ao) and body. See Figure 4 for further description of blood flow in tricuspid atresia. RA, right atrium, PA, pulmonary artery.

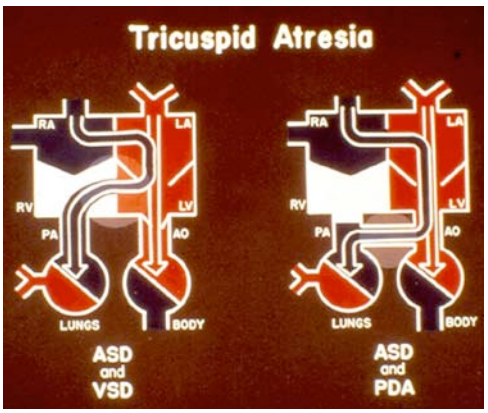

Figure 4. Box diagrams of tricuspid atresia: left panel with ventricular septal defect (VSD) and the right panel with patent ductus arteriosus (PDA). As seen in Figure 3, the right atrial (RA) blood exits into the left atrium (LA), mixes with pulmonary venous return then into the left ventricle (LV). In the presence of a VSD (left panel), shunting from the left to right (RV) ventricle takes place, thus perfusing the pulmonary artery (PA) and lungs. In the absence of VSD, but with PDA (right panel) left to right shunting occurs at the ductal level perfusing the PA and lungs. The presence of VSD, PDA or aorto-pulmonary collateral vessels is critical for patient survival.

of the great arteries, the atretic morphologic tricuspid valve is a left-sided atrioventricular valve and, therefore, in a physiological sense, it behaves as mitral (left or pulmonary venous atrial) obstruction. In other Type III and Type IV[53] patients, the systemic and pulmonary blood flows are determined by the size of the VSD and other associated defects.

Other Physiologic Principles

Arterial Desaturation. Because of complete admixture of the systemic, coronary, and pulmonary venous blood returns in the left atrium and left ventricle, systemic arterial desaturation is always present. The oxygen saturation is proportional to the magnitude of the pulmonary blood flow.[51,54] The data from our collection of patients are plotted in Figure 5; the pulmonary-to-systemic blood flow ratio (Qp:Qs) which represents the pulmonary blood flow has a curvilinear relationship with the arterial oxygen saturation. A Qp:Qs of 1.5 to 2.5 appears to result in an adequate oxygen saturation.[54] Further increase in Qp:Qs does not result in better oxygen saturation, but may subject the left ventricle to larger volume overloading and, therefore is not advisable.[54]

Pulmonary Blood Flow. The magnitude of pulmonary blood flow in an unoperated patient is dependent upon the degree of obstruction of the pulmonary outflow tract and patency of the ductus arteriosus. The pulmonary outflow obstruction is valvar or sub-valvar in Type II patients, and valvar, subvalvar, or at VSD level in Type I patients. In our own experience with several series of tricuspid atresia, we found the obstruction to be located most commonly at the VSD level.[39-42,45,46] When the VSD is large and non-restrictive and the pulmonary valve non-stenotic, the pulmonary flow is proportional to the pulmonary-to-systemic vascular resistance ratio. When a systemic-to-pulmonary artery shunt has been performed, the pulmonary blood flow is proportional to the size of the shunt.

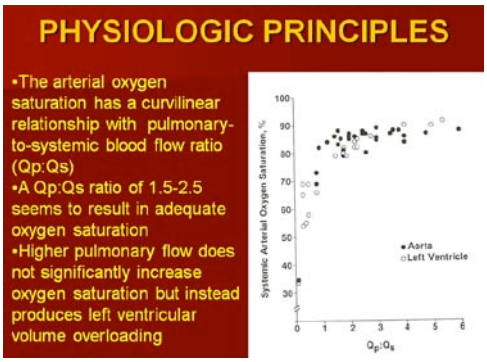

Figure 5. The systemic arterial saturations (left ventricular [LV] or aortic [Ao]) in patients with tricuspid atresia are plotted against the respective pulmonary to systemic blood flow ratios (Qp:Qs). Both Type I and Type II anatomy are included. Note curvilinear relationship between two parameters. At low Qp:Qs levels, a slight increase in Qp:Qs produces large increases in systemic O_2 saturation, whereas at higher Qp:Qs further increase does not produce significant increase in O_2 saturation. Ideal Qp:Qs appears to be between 1.5 to 2.5, giving O_2 saturations in low 80's. Aortic saturations are marked as solid circles and LV saturations as open circles.

Left Ventricular Volume Overloading. Because the entire systemic, coronary, and pulmonary venous blood returns are pumped by the left ventricle, the left ventricle has a greater volume overload than that in the normal heart. This volume overloading is further increased if the Qp:Qs is high either because of mild or no obstruction to pulmonary blood flow or because of large surgical shunts, either of which may result in heart failure. Normal left ventricular function is critical for successful Fontan-type of procedure and should be maintained within normal range. Several studies have shown that the left ventricular function tends to decrease with increasing age, Qp:Qs, and arterial desaturation.[55-57]

Size of the Interatrial Communication. The interatrial communication is usually a patent foramen ovale. Because of the obligatory shunting, this fetal pathway persists in the postnatal period; this is in part related to low left atrial pressure. However, the entire systemic venous return must pass through the patent foramen ovale and consequently, interatrial obstruction is expected, but very few patients with tricuspid atresia have clinically significant obstruction.[18] The right-to-left shunt occurs in late atrial diastole with augmentation during atrial systole ('a' wave).[54,58] A mean interatrial pressure difference greater than 5 mmHg is usually indicative of interatrial obstruction. A tall 'a' wave in the right atrium is also suggestive of interatrial obstruction.

Changing Hemodynamics. As the infant with tricuspid atresia grows and develops, several changes may take place. Closure of the ductus arteriosus occurring in the early neonatal period may result in severe hypoxemia. The size of the interatrial communication may diminish either in absolute terms or relative to the volume of the systemic venous return and cause systemic venous congestion and may require atrial septostomy. The ventricular septal defect may close spontaneously,[39-47] causing pulmonary oligemia and hypoxemia in Type I patients and subaortic obstruction in Type II patients. Such VSD closures occur over a period of months and years and are not germane to our discussion of tricuspid atresia in neonates. The reader is referred to other publications[26,41,42,46] for further discussion of this subject.

Clinical Features

The magnitude of pulmonary blood flow is the major determinant of clinical features in tricuspid atresia. An infant with markedly decreased pulmonary blood flow will present early in the neonatal period with severe cyanosis, hypoxemia, and acidosis. An infant with markedly increased pulmonary flow does not have significant cyanosis, but usually presents with signs of heart failure. Although there is some overlap, patients with decreased pulmonary flow usually belong to Type I (normally related great arteries) and those with increased pulmonary blood flow are usually Type II (transposition of the great arteries) and occasionally Type Ic. Approximately one-half of the patients with tricuspid atresia manifest symptoms on the first day of life and 80% would be symptomatic by the end of the first month of life.[18,59] Two modes of presentation are recognized:

1. Decreased pulmonary blood flow, and
2. Increased pulmonary blood flow.

Decreased Pulmonary Blood Flow

Infants with pulmonary oligemia present with symptoms of cyanosis within the first few days of life; 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 decreased. The majority of these infants belong to Type Ib. Patients with pulmonary atresia (Subgroup a) irrespective of the type 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.

Physical examination reveals central cyanosis, tachypnea or hyperpnea, normal pulses, prominent 'a' wave in the jugular venous pulse (if there is significant interatrial obstruction), and no hepatic enlargement (presystolic hepatic pulsations may be felt if there is severe interatrial obstruction). Quiet precordium and absence of thrills is usual. The second heart sound is usually single. A holosystolic type of 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 patent ductus arteriosus may be heard. Clinical signs of congestive heart failure are notably absent.

Increased Pulmonary Flow

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 week of life.[17] They are only minimally cyanotic, but manifest symptoms of dyspnea, fatigue, difficulty to feed, and marked perspiration. Recurrent respiratory tract infection and failure to thrive is another mode of presentation. The majority of these patients belong to Type IIc, although a small number of patients may be of Type Ic. The association of aortic coarctation with Type II patients has already been mentioned and coarctation, when present, makes them vulnerable to early cardiac failure.

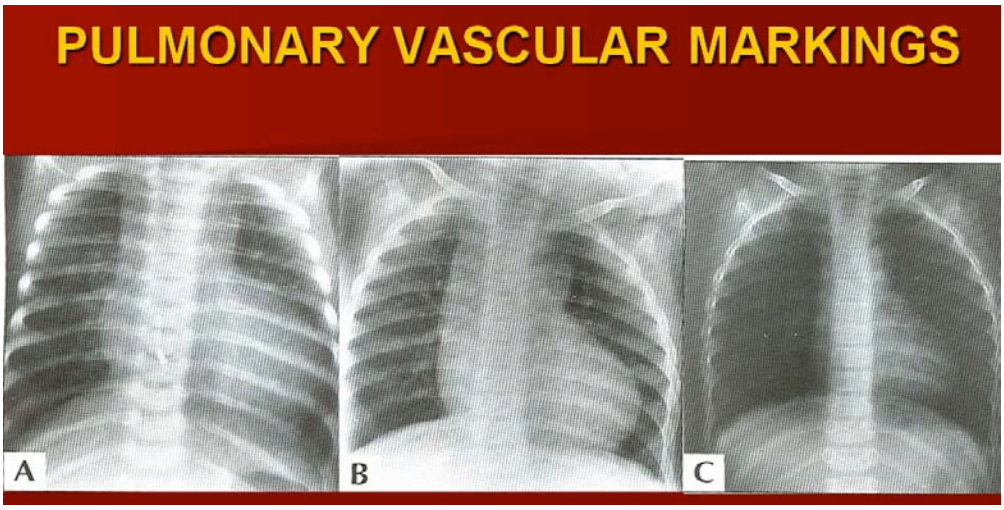

Figure 6. Chest roentgenograms (Posteroanterior views) of babies with tricuspid atresia showing enlarged cardiac size and increased pulmonary vascular markings (A); during follow-up, the size of the heart diminished and the pulmonary vascular markings decreased (B & C). Echocardiographic and angiographic studies demonstrated progressive decrease in the size of the ventricular septal defect.

Examination reveals tachypnea, tachycardia, decreased femoral pulses (when associated with aortic coarctation but without significant-sized patent ductus arteriosus), minimal cyanosis, prominent neck vein pulsations and hepatomegaly. Prominent 'a' waves in jugular veins and/or presystolic hepatic pulsations may be observed with associated interatrial obstruction. The precordial impulses are increased and hyperdynamic. 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 murmur may be heard. Clear-cut signs of congestive cardiac failure are usually present.

Noninvasive Evaluation

Chest Roentgenogram

Roentegenographic appearance is, by and large, dependent upon the total pulmonary blood flow (Figure 6C). In patients with diminished pulmonary flow, the heart size is either normal or minimally enlarged, whereas, in those with increased pulmonary blood flow, the heart size is moderately to severely enlarged. Several patterns of cardiac configuration, namely "characteristic" tricuspid atresia appearance,[60] *coeur en sabot* configuration,[61] "egg-shaped,"[28] "bell-shaped"[62] and square[20] heart have been described, but in the authors' experience and that of others,[28] there is no consistent pattern that would be diagnostic of tricuspid atresia. 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.

Right aortic arch may be present in approximately 8% of patients with tricuspid atresia[28] and is less common than that observed in patients with tetralogy of Fallot (25%) and truncus arteriosus (40%). An unusual contour of the left border of the heart suggestive of L-transposition may be seen in association with or confused with tricuspid atresia.[63]

The greatest use of the chest roentgenogram is its ability to categorize babies into those with decreased pulmonary vascular markings and into those with increased pulmonary vascular markings. Often, this is all that is necessary to make a correct diagnosis once a history, physical examination, and electrocardiogram have been obtained.[63]

Electrocardiogram

The electrocardiogram can be virtually diagnostic of tricuspid atresia in the neonate suspected to have a cyanotic congenital heart defect. Right atrial hypertrophy, an abnormal, superiorly oriented major QRS vector (so called left axis deviation) in the frontal plane, left ventricular hypertrophy, and diminished right ventricular forces (Figure 7) are characteristic findings.

Right atrial hypertrophy, manifested by tall, peaked P waves in excess of 2.5 mm, is present in the majority of the patients with tricuspid atresia.[64] Although it has been suggested that the amplitude of the P wave in lead II is directly proportional to the interatrial pressure difference and inversely proportional to the size of the interatrial communication, detailed analysis of these parameters did not suggest a consistent relationship.[18,65] A double peak, spike and dome configuration of the P wave, referred to as "P-tricuspidale"[64] may be present. The first taller peak is caused by the right atrial depolarization and the second smaller peak is presumed to be due to left atrial depoloarization.[64,66]

Abnormal, superiorly-oriented major QRS vector (ASV), more popularly called left axis deviation, between 0 to -90° in the frontal plane is present in the majority of the patients with tricuspid atresia. ASV is present in excess of 80% of patients with Type I anatomy (normally

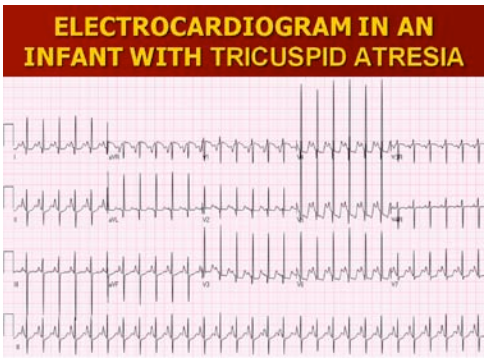

Figure 7. Twelve lead electrocardiogram showing abnormal, superiorly-oriented mean QRS vector in frontal plane (-45°, left axis deviation), left ventricular hypertrophy, diminished anterior (R waves in leads V1 and V2) and rightward (S waves in leads V5 and V6) forces. Prominent P waves, indicative of biatrial enlargement are also seen in several leads. This electrocardiogram is highly suggestive of tricuspid atresia.

related great arteries) while only less than 50% of patients with Type II and Type III anatomy show such a typical electrocardiographic pattern.[66] Normal (0 to +90°) or right axis deviation is present in a minority of patients and most of these patients belong to Type II or III anatomy. It has been suggested that the ASV may be related to destructive lesions in the left anterior bundle,[64] fibrosis of left bundle branch,[67] abnormal distribution of the conduction system (unusually long right bundle branch and origin of left bundle branch very close to the nodal-His bundle junction),[68-70] a small right ventricle or a large left ventricle.[65] Ventricular activation data from our group[66,71] suggested that this characteristic QRS pattern in tricuspid atresia is produced by interaction of several factors, the most important being the right-to-left ventricular disproportion and asymmetric distribution of the left ventricular mass favoring the superior wall.

Regardless of the frontal plane mean QRS vector orientation, electrocardiographic criteria for left ventricular hypertrophy are present in the vast majority of patients. This may be manifested by increased (above 95th percentile) S waves in right chest leads and R waves in left chest leads or by "adult progression" of the QRS in the chest leads in the neonates and infants. ST-T wave changes suggestive of left ventricular strain is present in 50% of patients.[66] The reason for left ventricular hypertrophy is the anatomic nature of the lesion, left ventricular volume overload and lack of opposition of the forces of left ventricular activation by the hypoplastic right ventricle. Rarely, biventricular hypertrophy may be present and the majority of these patients belong to Type II or III anatomy with good-sized right ventricle. Diminished R waves in right chest leads and S waves in left chest are related to right ventricular hypoplasia. Vectorcardiographic features closely resemble the scalar electrocardiogram, but vectorcardiography is no longer available for routine use.

Echocardiogram

M-mode echocardiography, while not diagnostic, is useful in evaluating the size of the left atrium and left ventricle and left ventricular function. Two-dimensional echocardiography, apart from showing enlarged right atrium, left atrium and left ventricle and a small right ventricle demonstrates the atretic tricuspid valve directly. In the most common muscular type, a dense band of echoes is seen at the site where tricuspid valve should be[55,72] and the anterior leaflet of the detectable atrioventricular valve is attached to the left side of interatrial septum (Figure 8). Apical and subcostal four-chambered views are best to demonstrate the anatomy. Atrial and ventricular septal defects can also be demonstrated by 2D 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. Suprasternal notch imaging will be of use in demonstrating coarctation of the aorta which is often seen in Type II 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, contrast study is not essential for making the diagnosis.

Doppler examination is useful in the evaluation of tricuspid atresia patients. The obligatory right-to-left shunt across the atrial septal defect can be demonstrated by placing pulsed Doppler sample volume on either side of the atrial septum and by color flow imaging (Figure 9). Left-to-right shunting across the VSD may also be

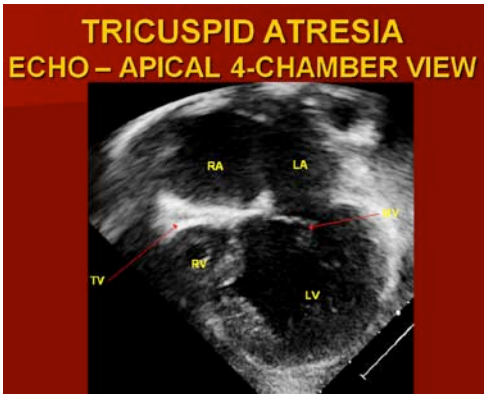

Figure 8. Apical four-chamber 2-dimensional echocardiographic view of an infant with tricuspid atresia showing enlarged left ventricle (LV), a very small right ventricle (RV), and a dense band of echoes at the site where the tricuspid valve echo should be (TV). Partially open mitral valve (MV) is also seen. Note the attachment of the anterior leaflet of the detectable atrio-ventricular valve to the left side of the interatrial septum. LA, Left atrium; RA, Right atrium.

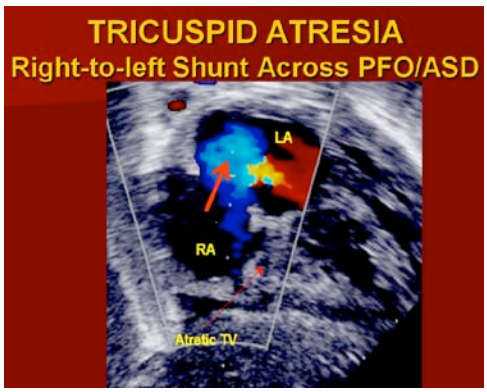

Figure 9. Selected video frame from an infant with tricuspid atresia demonstrating right-to-left (blue flow) shunt (thick arrow) across the interatrial communication in a subcostal four-chamber view. Atretic tricuspid valve (thin arrow) is seen. LA, Left atrium; RA, Right atrium.

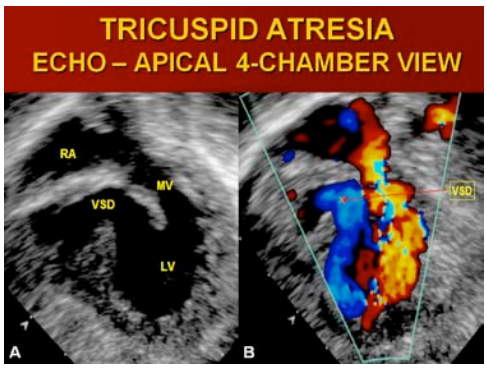

Figure 10. Selected video frames from an infant with tricuspid atresia of modified apical four-chamber 2-dimensional echocardiographic views without (A) and with color-flow Doppler (B) illustrating enlarged left ventricle (LV), small right ventricle (not labeled), atretic tricuspid valve (not labeled), open mitral valve (MV) and a ventricular septal defect (VSD) with shunting across it.

demonstrated by Doppler (Figure 10). In Type I (normally related great arteries) patients, the VSD peak Doppler velocity is helpful in estimating the size of the VSD; the higher the velocity, the smaller is the VSD. Right ventricular and pulmonary arterial pressure may also be estimated using modified Bernoulli equation:

$$\text{RV/PA systolic pressure} = \text{systolic BP} - 4V^2$$

Where, RV is right ventricle, PA is pulmonary artery, BP is arm blood pressure and V is VSD peak Doppler velocity.

In the presence of pulmonary hypertension or severe infundibular or valvar pulmonary stenosis, the VSD Doppler velocities are not indicative of the size of the VSD. In Type II (D-transposition) patients, high VSD velocity is suggestive of subaortic obstruction.

Interrogation of right ventricular outflow tract in Type I patients and pulmonary artery region in Type II patients may reveal pulmonary or subpulmonary stenosis; higher the velocity, more severe is the obstruction. Doppler evaluation of descending aortic flow is helpful in demonstrating aortic coarctation.

In summary, delineation of the majority of anatomic and physiologic issues related to tricuspid atresia is feasible by M-mode, 2-dimensional and Doppler (pulsed, continuous wave and color) echocardiography, and when indicated, contrast echocardiography.

Other Laboratory Studies

Pulse oxymetry and blood gas values are useful in quantitating the degree of hypoxemia, thereby indicating the severity of pulmonary oligemia. Hemoglobin and hematocrit values are not particularly useful in the neonate, but the degree of polycythemia is useful in estimating the severity of hypoxemia at a later age.[73]

Cardiac Catheretization

The diagnosis of tricuspid atresia based on clinical, electrocardiographic and echocardiographic features is relatively simple, and cardiac catheterization and selective cineangiography, rarely, if ever, are essential for establishing the diagnosis.[54] Even neonates with significant arterial desaturation need not undergo cardiac catheterization and selective cineangiography; the diagnosis of tricuspid atresia is usually made on the basis of clinical and non-invasive evaluation, particularly echo-Doppler studies. Catheterization may be indicated:

1) prior to bidirectional Glenn procedure and Fontan correction, particularly to define the pulmonary artery anatomy and
2) if catheter-based atrial septostomy is required.

Since the neonates do not usually require cardiac catheterization and selective cineangiography, these will not be discussed; the interested reader is referred to elsewhere.[54,74,75]

Differential Diagnosis

Differential diagnostic considerations differ with the mode of presentation: a) moderate to severely cyanotic infants with decreased pulmonary vascular markings on chest roentgenogram, and b) mildly cyanotic infants with increased pulmonary vascular markings and with or without signs of congestive heart failure.

Decreased Pulmonary Blood Flow

Once decreased pulmonary blood flow is recognized on the chest film, several possibilities, as listed in Table II, should be considered. Most often, they can be differentiated with the help of an electrocardiogram (Figure 11).[3]

Echocardiography and/or cineangiography are confirmatory.

Tetralogy of Fallot (including VSD with Pulmonary Atresia). The frontal plane mean QRS vector is between 90° and 180° in this group and right ventricular hypertrophy is present. Echocardiograms demonstrate a large right ventricle, a large aorta that overrides the interventricular septum, a large subaortic VSD and increased Doppler flow velocity across the pulmonary outflow tract. Angiographic features are characteristic for this anomaly.

Pulmonary Atresia (or Stenosis) with Intact Ventricular Septum and Hypoplastic Right Ventricle. The mean frontal plane vector is between 0° to 90° without right ventricular hypertrophy; left ventricular hypertrophy may be present and right ventricular forces may be decreased. Echocardiogram shows a large left ventricle, a hypoplastic right ventricle and small but demonstrable tricuspid valve leaflets. Angiocardiography will confirm the diagnosis.

Tricuspid Atresia. These patients will not only have an abnormal, superiorly-oriented QRS vector (0° to -90°) on the frontal plane, but will also have left ventricular hypertrophy and decreased right ventricular forces.

Complex Pulmonary Stenosis. This group includes several defects, namely, single (double-inlet left) ventricle, double outlet right ventricle, transposition of the great arteries with VSD, ventricular inversion and others, all associated with severe pulmonary stenosis or atresia. The electrocardiographic mean frontal plane vector and ventricular hypertrophy patterns vary markedly. Echocardiography and/or angiography are often necessary for accurate diagnosis.

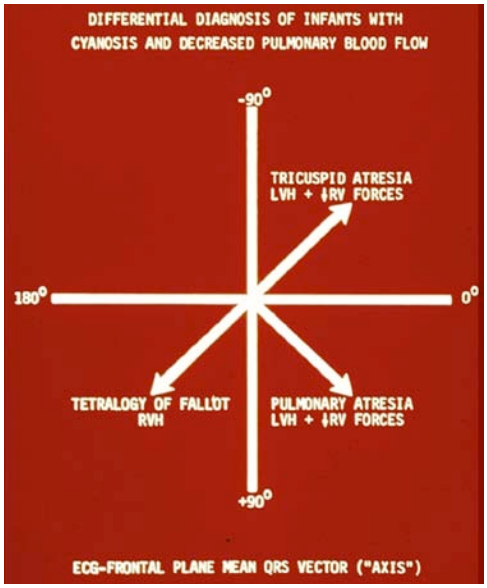

Figure 11. The figure illustrates the usefulness of frontal plane mean QRS vector (axis) in the differential diagnosis of cyanotic heart defects with decreased pulmonary flow. An axis between 0 and –90⁰ is suggestive of tricuspid atresia, an axis between 0 and +90⁰ is indicative of pulmonary atresia with intact ventricular septum and an axis between +90⁰ and ±180⁰ is associated with tetralogy of Fallot. The associated ECG abnormalities are shown in the respective quadrant. Cyanotic infants with more complex heart defects with severe pulmonary stenosis/atresia may fall in any quadrant.

Increased Pulmonary Blood Flow

The differential diagnostic considerations are also listed in Table II. Although the characteristic electrocardiographic pattern (abnormal, superior vector or "left axis deviation") is helpful, it is not present in all cases of tricuspid atresia with transposition. Furthermore, some of the conditions listed in Table IIB also have similar displacement of mean frontal plane vector. Often, echocardiograms and occasionally, angiocardiograms are necessary for final diagnosis.

Management

Physiologically "corrective" surgery for tricuspid atresia[77,78] and its modifications[79-81] have improved the prognosis of patients with tricuspid atresia. Such physiologic correction is usually performed in patients older than 2 years, at an approximate weight of 15 Kg. As stated previously, most tricuspid atresia patients manifest 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 pressure and preserved left ventricular function) to normal such that a "corrective" procedure could be performed at an appropriate age. Keeping the above objective in mind, the management plan may be discussed under the following headings:

1) medical management at the time of initial presentation,
2) palliative treatment of specific physiologic abnormalities,
3) medical management following palliative surgery,
4) physiologically "corrective" surgery, and
5) follow-up after corrective operation.

Items 3 to 5 are not relevant to a chapter on tricuspid atresia in neonates and, therefore, will not be detailed.

Medical Management at the Time of Initial Presentation

The need for prompt identification and rapid transfer of a cyanotic/distressed neonate with suspected serious heart disease to a regional pediatric cardiology center is well recognized. During the process of identification, transfer to a pediatric cardiology center, initial work-up, and palliative surgery, as well as following surgery, neutral thermal environment, normal acid-base status, normoglycemia, and normocalcemia should be maintained by appropriate monitoring and correction if needed.[4] No more than 0.4 $F1O_2$ is necessary unless pulmonary parenchymal disease is present.

Infants with low arterial PO_2 and decreased oxygen saturation may be ductal-dependent; therefore, the ductus should be kept open by intravenous administration of prostaglandin E_1 (PGE_1).[4,82] The ductal dilating effect of this drug results in an increase in pulmonary blood flow, thereby improving oxygenation and reversing the metabolic acidosis so that further diagnostic studies and surgical intervention can be performed with relative safety. Current suggestions are for intravenous infusion of PGE_1 at a dose of 0.05 to 0.1 µg per kilogram of body weight per minute. We usually begin with a dose of 0.05 µg/kg/min and reduce the rate of infusion, provided the desired oxygen saturation levels are achieved; this has been most helpful in reducing the incidence and severity of some of the drug's bothersome side effects, namely, apnea and hyperpyrexia. PGE_1 infusion rate may be increased if there is no increase in PO_2.

The occasional infant that presents with signs of congestive heart failure (more common in Type II patients) should be treated with routine anti-congestive measures.[83] Patients with associated severe coarctation of the aorta may also be

Table II.
Differential Diagnosis of Tricuspid Atresia in the Neonate

A. Decreased Pulmonary Blood Flow	
	1. Tetralogy of Fallot including pulmonary atresia with ventricular septal defect 2. Pulmonary atresia or severe stenosis with intact ventricular septum 3. Tricuspid atresia 4. Complex cardiac defects with severe pulmonary stenosis or atresia
B. Increased Pulmonary Blood Flow	
	1. D-Transposition of the great arteries with large ventricular septal defect 2. Coarctation of the aorta with ventricular septal defect 3. Multiple left-to-right shunts (ventricular septal defect, common atrioventricular canal and patent ductus arteriosus) 4. Single ventricle, double outlet right ventricle and other complex cardiac defects without pulmonary stenosis 5. Total anomalous pulmonary venous connection without obstruction 6. Hypoplastic Left Heart Syndrome

helped with PGE_1 infusion; this time the ductal dilatation improves systemic perfusion. This should be followed by surgical relief of coarctation. Alternatively, balloon angioplasty may be utilized to relieve the aortic obstruction.[84-86]

Palliative Treatment of Specific Physiologic Abnormalities

The palliation of patients with tricuspid atresia would largely depend upon the hemodynamic abnormality produced by the basic lesion and associated cardiac defects. These abnormalities may be broadly grouped[4,50] into:

1) decreased pulmonary blood flow
2) increased pulmonary blood flow, and
3) intracardiac obstruction.

Decreased Pulmonary Blood Flow. Since the description of subclavian artery-to-ipsilateral pulmonary artery anastomosis (Figure 12) in 1945 by Blalock and Taussig,[87] several other types of operations/procedures have been devised to improve the pulmonary blood flow. These include other types of systemic-pulmonary artery shunts, namely: the Potts anastomosis (descending aorta-to-left pulmonary artery shunt), ascending aorta-to-main pulmonary artery anastomosis (central shunt), Waterston-Cooley shunt (ascending aorta-to-right pulmonary artery anastomosis), aorta-to-pulmonary artery Gore-Tex shunt, and Gore-Tex interposition graft between the subclavian artery and the ipsilateral pulmonary artery, superior vena cava-to-right pulmonary artery anastomosis (Glenn Procedure), formalin infiltration or stenting the ductus arteriosus, and enlarging the VSD. Systemic-pulmonary artery shunts are most commonly used in the palliation of pulmonary oligemia. Because of the problems associated with central shunts, the Blalock-Taussig type of shunt is preferred. At present, a modified Blalock-Taussig shunt with a Gore-Tex graft (Figure 13A) interposed between the subclavian artery and the ipsilateral pulmonary artery[88] appears to be the first choice in most institutions for palliation of the neonate and young infant with pulmonary oligemia.

Enlargement of the VSD and/or resection of the right ventricular outflow tract stenosis has been performed and recommended by Annecchino and his colleagues[89] as a palliative procedure to augment the pulmonary blood flow. This is an ingenious approach and attacks the site of obstruction rather than bypassing it. However, it is an open heart procedure and may not be feasible or necessary in the neonatal period.[50] Placement of a stent in the ductus, to keep it open to provide pulmonary flow is an attractive option,[90,91] but because of limited experience and the technically demanding nature of the procedure, it currently is not a therapeutic procedure of choice. If the predominant obstruction is at the pulmonary valve level, balloon pulmonary valvuloplasty may be considered.[92]

In conclusion, despite the availability of many types of palliative procedures to increase pulmonary blood flow, most of them are either not effective or, if effective, may produce serious complications to deter from performing a successful Fontan-Kreutzer procedure subsequently. Blalock-Taussig anastomosis or one of its modified versions is the preferred procedure and has the least number of long-term complications, but at the same time, preserves suitable anatomy for subsequent corrective procedures. Therefore, it is recommended as the procedure of choice for palliation of tricuspid atresia patients with decreased pulmonary blood flow.

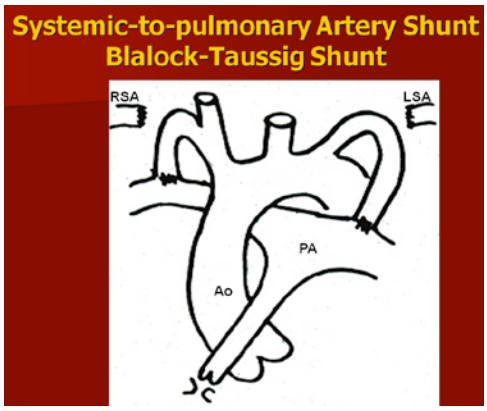

Figure 12. Diagrammatic portrayal of classic Blalock-Taussig shunt with subclavian artery-to-ipsilateral pulmonary artery anastomosis. The disconnected right (RSA) and left (LSA) subclavian arteries are shown. Ao, Aorta; PA, Pulmonary artery.

Increased Pulmonary Blood Flow. Infants with a modest increase in pulmonary blood flow do not have any significant symptomatology and, indeed, are less cyanotic than the pulmonary oligemic patients. Markedly increased pulmonary blood flow, however, can produce congestive heart failure. Only Type Ic and Type IIc patients, i.e., without associated pulmonary stenosis, will fall into the category of pulmonary plethora. A majority of these patients will have Type II anatomy and will usually manifest during early infancy.

In Type I patients, aggressive anticongestive measures should be promptly instituted. The natural history of the VSD has been well documented in this group;[39-46] the VSD becomes smaller and patients with pulmonary plethora will, in due course, develop pulmonary oligemia (Figure 6), requiring palliative surgical shunts. These patients can also develop right ventricular outflow tract obstruction with resultant decrease in pulmonary blood flow. Therefore, it is generally recommended that pulmonary artery banding not be performed in this group of patients. Among our 40 consecutive patients with tricuspid atresia,[46,50] only two with Type I anatomy required pulmonary artery banding and there are only a few cases reported in the literature that required pulmonary artery banding. If optimal anticongestive therapy with some time delay does not produce adequate relief of symptoms,[50] pulmonary artery banding should be considered in Type I patients; perhaps a serious consideration for using absorbable band material[93-95] should be given. Bonnet et al. used absorbable pulmonary artery band for palliation in such infants.[94] 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 that might have been associated with a conventional non-absorbable band. This is an innovative approach, although it is likely to be helpful in a limited number of patients.[95] In those that did not have pulmonary artery banding performed, careful follow-up studies with measurement of pulmonary artery pressure and appropriate treatment are necessary to prevent pulmonary vascular obstructive disease.

In Type II patients, banding of the pulmonary artery (Figure 13B) should be performed once the infant is stabilized with anticongestive therapy. If there is associated coarctation of the aorta, or aortic arch interruption or hypoplasia, adequate relief of the aortic obstruction should be provided concurrent with pulmonary artery banding, and the patent ductus arteriosus should be ligated, if present. The importance of PGE_1 administration in temporarily relieving aortic obstruction and thereby control congestive heart failure has already been alluded to. The role of balloon dilatation angioplasty of the coarctation[84-86] in these complicated lesions has not yet been completely delineated. Because of higher risk for poor outcome in patients with transposition and those requiring pulmonary artery banding and/or aortic arch repair,[96] early, adequate and appropriate interventions are desirable.

Intracardiac Obstruction. Intracardiac obstruction can occur at two different levels, namely, patent foramen ovale and VSD.

Interatrial Obstruction. Since the entire systemic venous return must egress through the patent foramen ovale, it should be of adequate size to accommodate it. A mean atrial pressure difference of 5 mmHg or more with very prominent 'a' waves (15 to 20 mmHg) in the right atrium is generally considered to represent obstructed interatrial septum.[50] Balloon atrial septostomy;[97] if unsuccessful, blade atrial septostomy;[98,99] and; rarely surgical atrial septostomy may become necessary to relieve the obstruction. Significant interatrial obstruction requiring atrial septostomy in the neonate is rare and unusual, although this can be a significant problem later in infancy.[41]

Interventricular Obstruction. Spontaneous closure of the VSD causing severe pulmonary oligemia in Type I patients and subaortic obstruction in Type II patients can occur;[39-47] this usually takes months to years to develop. For further discussion of this subject, the reader is referred elsewhere.[41,42,46]

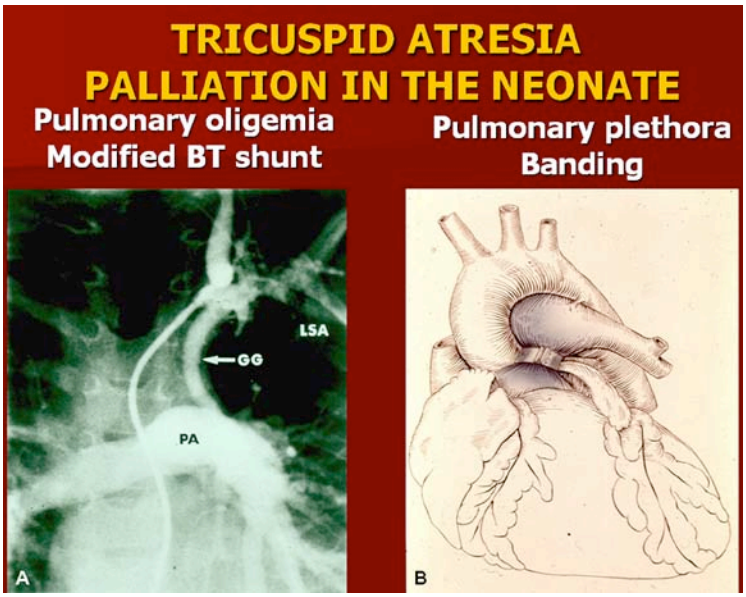

Figure 13. Palliation of tricuspid atresia patients. Patients with pulmonary oligemia with markedly decreased O_2 saturation may be palliated by a modified Blalock-Taussig shunt, connecting the subclavian artery to the ipsilateral pulmonary artery with a Gore-Tex graft (GG); the left panel (A) demonstrates GG opacified on left subclavian artery (LSA) cineangiogram with excellent opacification of the pulmonary artery (PA). In patients with pulmonary oligemia, but with O_2 saturations in low 80s may be followed clinically without any intervention. Patients with markedly increased pulmonary blood flow and severe congestive heart failure may require banding of the pulmonary artery, as shown in right panel (B).

Management Beyond Neonatal Period

Detailed discussion of this subject is beyond the scope of this review and the reader is referred to other publications[50,76,100,101] for further treatment of this subject. Since the description by Fontan[77] and Kreutzer,[78] the procedure has evolved[79-81] and continues to evolve.[76,100,101] At the time of this writing, most authorities seem to prefer total cavopulmonary connection,[79] performed as a two-staged procedure; bidirectional Glenn[102] initially followed by Fontan conversion either by an intra-atrial lateral tunnel or an extracardiac nonvalved conduit diversion[103] of the inferior vena caval and hepatic blood into the pulmonary circuit a year or so later. In the presence of risk factors,[104] fenestration[105,106] may be performed which could be closed either in the immediate post-operative period[106] or at a later time by transcatheter methodology.[105,107]

Summary of Surgical Management

Physiologically corrective surgery is feasible in most patients with tricuspid atresia. The age and size of the patient at presentation and anatomic and physiologic substrate dictate the type of palliative/corrective procedures. In the neonate and young infant (≤ 3 months) with pulmonary oligemia, a modified Blalock-Taussig shunt is the procedure of choice. Some infants with O_2 saturations in low 80s may not require any intervention. Between 3 to 12 months, a modified Blalock-Taussig shunt and a bidirectional Glenn procedure are the available options; we would prefer bidirectional Glenn. Between 12 to 24 months, bidirectional Glenn is preferred. Beyond 2-years-of-age, total cavopulmonary connection with or without fenestration, depending upon the presence of risk factors, appears to be the choice, although some authorities stage the Fontan even at this age. Early pulmonary artery banding, relief of aortic arch obstruction and relieving or bypassing subaortic obstruction should be incorporated into the treatment plan in patients with tricuspid atresia and transposition of the great arteries.

Prognosis

Untreated, the prognosis of live born infants with tricuspid atresia is poor; only 10 to 20% may survive their first birthday.[17,18] Palliative surgery to normalize the pulmonary blood flow has markedly improved the survival rate. But, as one can see from survival data from several large studies,[26,59,96] there is still considerable early mortality. Because of recent improvement in surgical mortality for the palliative surgery and advances in neonatal care, the initial mortality should decrease. Introduction of physiologically "corrective" surgery in the early 1970s has, to some degree, improved the second bout of mortality seen in children beyond 15-years-of-age. Because of this improved prognosis, each neonate with tricuspid atresia should be offered aggressive medical and surgical therapy.

Summary/Conclusion

Tricuspid atresia is the third most common cyanotic congenital heart defect. There are significant variations in the morphology of the atretic tricuspid valve, associated cardiac defects and physiology, resulting in different clinical presentations. The diagnosis is relatively simple and can often be made by careful review of clinical features and simple laboratory studies (chest roentgenogram and electrocardiogram) which can be confirmed by echocardiography. Aggressive management to normalize the pulmonary blood flow and correct physiologically important associated defects (for example coarctation of the aorta) should be undertaken at the time of presentation. Follow-up and treatment plans should strive to maintain or normalize cardiac structures and function (pulmonary artery anatomy and pressure, and left ventricular function). Finally, performing modified, staged Fontan-Kreutzer surgery prior to deterioration of the left ventricular function should markedly improve the prognosis for tricuspid atresia patients.

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Corresponding Author

P. Syamasundar Rao, MD
Professor of Pediatrics and Medicine
Emeritus Chief of Pediatric Cardiology
The University of Texas/Houston Medical School
Children's Memorial Hermann Hospital
Houston, Texas

Address for correspondence:
Dr. P. Syamasundar Rao
Professor and Director,
Division of Pediatric Cardiology,
The University of Texas/Houston Medical School
6431 Fannin, MSB 3.130
Houston, TX 77030, USA
Phone: 713.500.5738; Fax: 713.500.5751

P.Syamasundar.Rao@uth.tmc.edu

Srilatha Alapati, MD
Interventional Pediatric Cardiology Fellow
University of Texas/Houston Medical School
Children's Memorial Hermann Hospital
Houston, Texas

Medical News, Products and Information

Women & Infants' Physician Named 2012 Legend of Neonatology

William Oh, MD, former Chief of Pediatrics at Women & Infants Hospital of Rhode Island and The Warren Alpert Medical School of Brown University, was recently inducted into the *Legends of Neonatology Hall of Fame* which was established in 2007. Dr. Oh was one of two physicians inducted this year at the *NEO* meeting in February 2012 for contributions to the care of the critically ill neonate.

In April, Dr. Oh also received another prestigious award, the *Maureen Andrews Mentorship Award*, from the Society for Pediatric Research, during its annual meeting in Boston, in recognition of his mentoring career. In the past four decades, Dr. Oh has trained more than 80 neonatologists, who are now leaders in their fields all over the world.

Dr. Oh is one of the founders of the field of neonatal medicine and has been a leader in teaching about metabolism, minerals, and fluids and electrolytes in the newborn infant.

"This is such an honor for Dr. Oh and his colleagues at Women & Infants and Brown," said Constance A. Howes, President & CEO of Women & Infants. "The contributions that Dr. Oh has made to the field of neonatology are extraordinary. He has impacted the care and caring of some of the tiniest, frailest babies and has paved the way for incredible discoveries and improvements in the care that we provide here and globally."

Originally trained in the Philippines where he received his medical degree, Dr. Oh came to the US in 1958, to do a pediatric residency at Michael Reese Hospital in Chicago, where he later became chief resident and a research fellow in neonatology. From 1964 to 1966, he initiated a series of research projects at the Karolinska Institute in Stockholm that resulted in one of the first series of papers to examine neonatal blood pressure, neonatal blood volume, neonatal hemodynamics, and neonatal renal function.

Dr. Oh became Director of Neonatology at Michael Reese Hospital in 1966, and in 1969 joined the faculty as Chief of Neonatology at Harbor General Hospital in California until 1974. In 1975 Dr. Oh left California to become Pediatrician-in-Chief of Women & Infants Hospital and Professor of Pediatrics and Obstetrics at Brown University, where he was appointed Chairman of the Department of Pediatrics in 1989. During this highly productive part of his career, Dr. Oh published virtually non-stop in a number of areas of neonatal medicine.

He continued his efforts at understanding neonatal blood pressure, the role of acid-base balance upon abnormal fetal heart rate patterns and neonatal well-being, the effects of insensible water loss upon neonatal metabolism, nutritional well-being in neonates, neonatal glucose metabolism, intrauterine growth retardation, neonatal renal function, bilirubin toxicity, and many other issues.

Increasingly, Dr. Oh has become interested in long-term neurodevelopmental outcome following neonatal intensive care unit (NICU) hospitalization and has been a leading figure in the NICHD Neonatal Network. He has won numerous major awards and honors, including the Apgar Award of the American Academy of Pediatrics. Dr. Oh has long been focused on one of the key areas of modern medicine, namely improving outcomes for neonates, and has contributed as much as any living neonatologist in that regard.

In 2009, Women & Infants opened the country's largest, single-family room neonatal intensive care unit. See a related article the February 2010 issue of *Neonatology Today*, by James F. Padbury, MD and Barry M. Lester, PhD, entitled, "Millennium Neonatology: Building for the Future." www.neonatologytoday.net/newsletters/nt-feb10.pdf.

Neonatal Lung Function Deficits in Children Who Develop Asthma

Children who develop asthma by age seven have deficits in lung function and increased bronchial responsiveness as neonates, a new study from researchers in Denmark suggests.

"Previous research on the relationship between neonatal lung function and the development of asthma has been conflicting," said lead author Hans Bisgaard, MD, DMSci, Professor of Pediatrics at the University of Copenhagen and head of the Danish Pediatric Asthma Centre. "Our study shows that children with asthma by age seven already had significant airflow deficits and increased bronchial responsiveness as neonates. Lung function deficits also progressed throughout childhood in our study, suggesting a potential opportunity for early intervention."

The findings were published online ahead of print publication in the American Thoracic Society's *American Journal of Respiratory and Critical Care Medicine*.

The prospective study enrolled a birth cohort of 411 at-risk children of asthmatic mothers. Spirometry was performed at one month in 403 (98%) children and again at age seven in 317 (77%).

Significant neonatal airflow deficits, as measured by forced expiratory flow at 50% of vital capacity and forced expiratory volume after 0.5 seconds, were observed among the 14% of children who developed asthma by age seven. Bronchial responsiveness to methacholine, which provokes narrowing of the airways, was also significantly associated with the development of asthma. Neonatal airway reactivity was a stronger predictor of asthma than neonatal lung function.

"We found that approximately 40% of the airflow deficit that was associated with asthma in our study was present at birth, while 60% developed through early childhood along with the disease," noted Dr. Bisgaard. "This indicates that both prenatal and early childhood mechanisms are potential intervention targets for the prevention of asthma."

The study used a homogenous study sample, which might limit extrapolation of the results to other populations.

"It seems that lung function changes associated with asthma occur very early in life and maybe even before birth," concluded Dr. Bisgaard. "This may explain the lack of effect from early intervention with inhaled corticosteroids and should direct research into the pathogenesis and prevention of asthma towards the earliest phases of life."

To read the article in full, please visit: www.thoracic.org//media/press-releases/resources/bisgaard.pdf.

The American Journal of Respiratory and Critical Care Medicine (AJRRCM) is a peer-reviewed journal published by the American Thoracic Society. It aims to publish the most innovative science and the highest quality reviews, practice guidelines and statements in the pulmonary, critical care and sleep-related fields.

Global Neonatology Today Monthly Column - Tracking the MGDs: What is the Status of Poverty in 2012

By Dharmapuri Vidyasagar, MD, FAAP, FCCM

Now that the target date of 2015 for reaching the United Nation's MDGs (Millennium Development Goals) is less than 3 years away, major funding agencies and stakeholders are continuously monitoring the progress of all MDGs.

Here are some of the findings of the International Monitory Fund (IMF) and the World Bank (WB) regarding the MDG #1 - "Eradicate Extreme Poverty."

According to the IMF and WB, the world is on track to reduce by half the number of people living in extreme poverty (i.e living on less than $1.25/day). According to the report, by 2015 eight-hundred-and-eighty-three million people will live in poverty. This is less than the 918 million that had been projected previously. The observed decline is mainly because of the significant changes that occurred in China and India.

Overall, there was a large global reduction of poverty between 1990-2011, and it is projected to decrease further by 2015. In 2005, 42% of the world's population (1.8 billion) were living on less than $1.25/day. That is expected to drop to 25% (1.3 billion) by 2015.

Looking at the progress in different regions and countries, it is clear that most of the progress has been made in East Asia, particularly in China. In 1990, 60% of the population (683 million) of China continued to earn less than $1.25/day; it dropped to 16% in 2005, and is expected to drop to 4.8% by the year 2015. By then, only 66 million people in China will remain earning below $1.25/day. This very significant achievement is well beyond the set target.

It is interesting to note that in 1990 the poverty rate in India was lower than in China (52% vs 60%). Although there is an overall decrease in percentage and the number of people living on less than $1.25/day, the decrease in India is at a much slower rate than that is seen in China. In India, it decreased from 52% in 1990 to 42% in 2005, while during that same period in China it dropped to 16% in 2005.

"Overall, in 2015, 25% of the world's population (1.38 billion) is expected to remain in poverty earning less than $1.25/day."

The trends in Sub-Saharan Africa also show a decrease, but at a much slower rate; it dropped from a high of 57% to 51% in 2005, and is expected to drop to 36%, but still remain higher than in East Asia and South Asia.

Overall, in 2015, 25% of the world's population (1.38 billion) is expected to remain in poverty earning less than $1.25/day.

Although these dropping numbers of extreme poverty are very encouraging, the poorest countries remain poor. At the current rate, by 2015, one billion people will be still extremely poor by the standards of middle and high income countries. The $1.25/day poverty line is the average for the world's poorest 10-20 countries. A higher line of $2.00/day, the median poverty line for developing countries, shows less progress.

"Having 22% of people in developing countries still living on less than $1.25 a day and 43% with less than $2.00 a day is intolerable. We need to increase our efforts..., we need to continue attacking poverty on many fronts… particularly in low-income countries," says Jaime Saavedra, Director of the World Bank's Poverty Reduction and Equity Group (Jan. 2012).

The Clock is Ticking !!!

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Dharmapuri Vidyasagar, MD, FAAP, FCCM
University of Illinois at Chicago
Professor Emeritus Pediatrics
Division of Neonatology
Phone: +312.996.4185
Fax: 312.413.7901
dvsagarmd@yahoo.com

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Corporate Offices:
8100 Leaward Way, Nehalem, OR 97131 USA

Mailing Address:
PO Box 444, Manzanita, OR 97131 USA

Tel: +1.301.279.2005; Fax: +1.240.465.0692
www.NeonatologyToday.net

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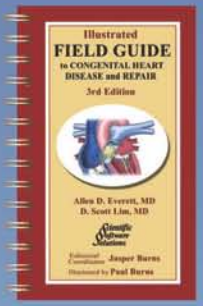

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