Thirty two years old woman during late outcome Fontan procedure consequence of tricuspid atresia

Background

The malformation of tricuspid atresia consists of a complete agenesis of the tricuspid valve with an absence of a direct communication between the right atrium and right ventricle.

Tricuspid atresia is the third most common form of cyanotic congenital heart disease. It is also the most common cause of cyanosis with left ventricular hypertrophy.

The natural history of this condition is such that, without surgical intervention, only one third of patients survive to age 1 year and only 10% live to age 10 years.(1)

The Fontan procedure, which was first performed in 1968 and then described in 1971, has changed the natural history dramatically and allowed survival into the third and fourth decades of life. (2)

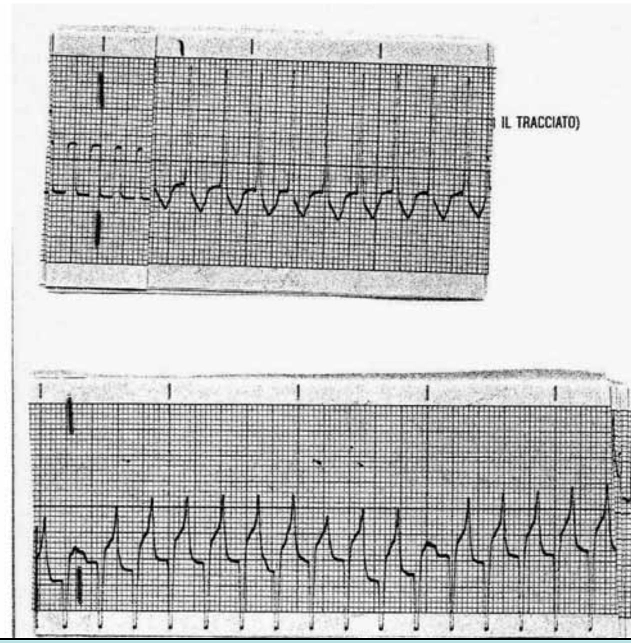
1. ***Keith JD, Rowe RD, Vlad P. Tricuspid Atresia. New York, NY: Macmillan; 1958.***
2. ***Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax. May 1971; 26: 240-248.***

BS, feminine 32 yo. Date: April 30, 2012. Late post operation Fontan Procedure for Tricuspid atresia

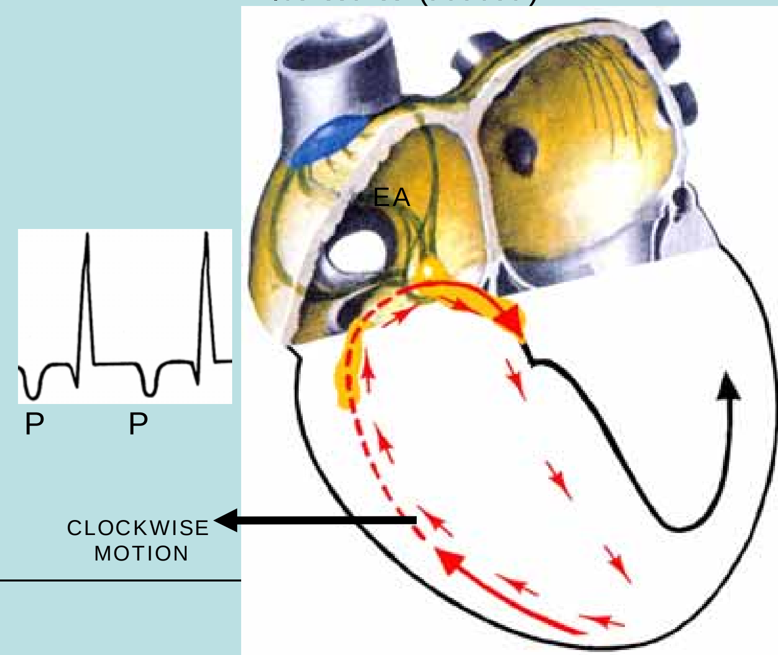
ECG diagnosis: Ectopic atrial rhythm P axis -60º(P wave isodipahsic in AVR) Low right atrial rhythm. Biatrial enlargement: Peaked P and broad notched (P duration 120ms) Deep prominent negative final component in V1 and positive P polarity in V2 in II Normal PA and extensive VSD. Prolonged PR interval: first degree AV block. QRS axis near 0º. Left ventricular enlargement/hypetrophy with LV strain pattern

Rarely adults with adequate RV have QRS axis between 0º and +90º. Absence of Left Anterior Fascicular Block pattern is suggestive of Group II of TA: TA with D-transposition without pulmonary stenosis. Counterclockwise rotation of the QRS loop in FP without LAFB-like pattern.

Tachycardial event



PAROXYSMAL SUPRAVENTRICULAR TACHYCARDIAS BY RECIPROCAL MACRO-REENTRY IN WPW (AVRT)



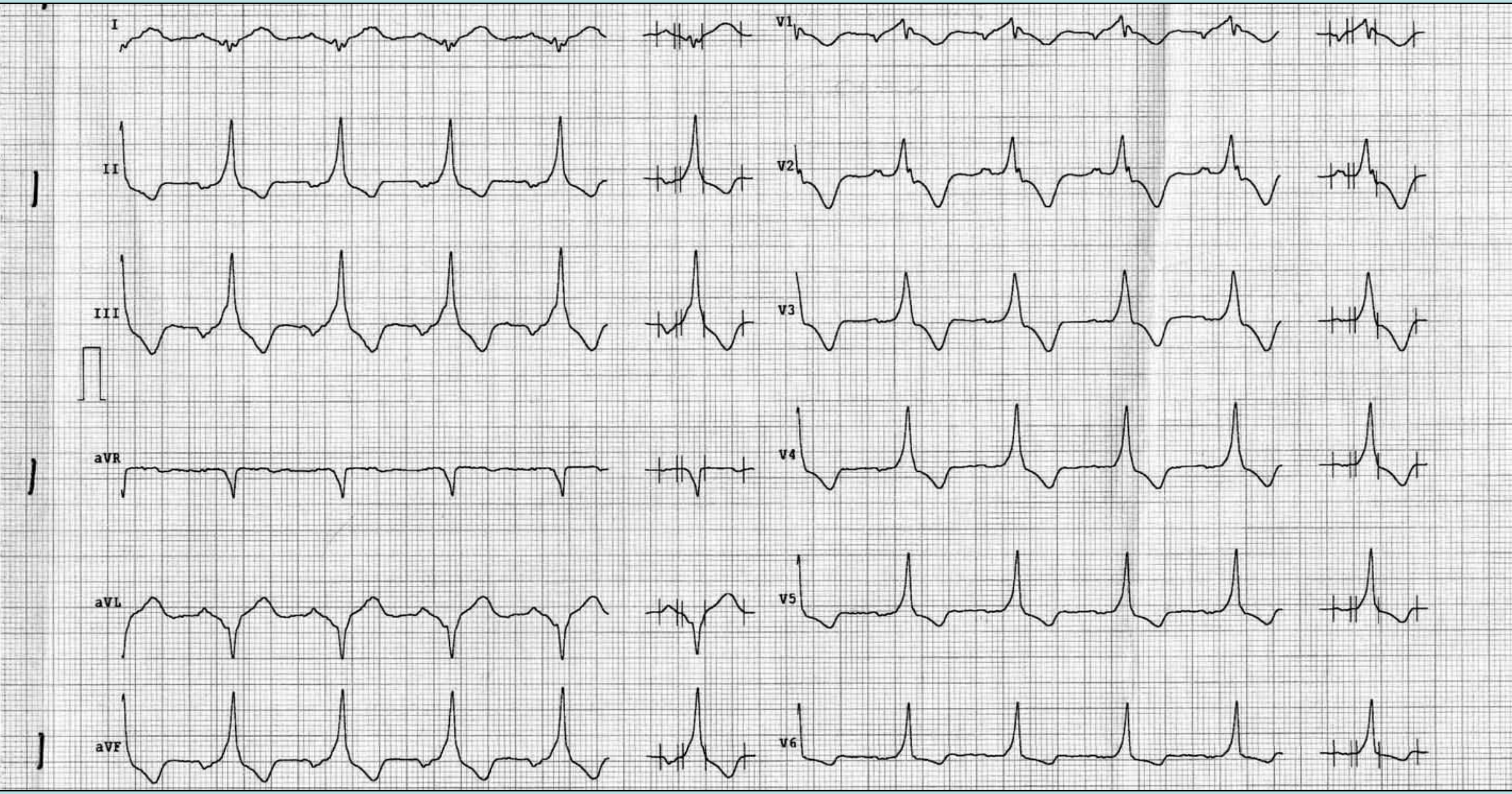
Narrow QRS Paroxysmal Tachycardia (PSVT): which one?

Answer: Orthodromic Circus Movement Tachycardia. It is usually initiated by an atrial premature beat and supported by an atrioventricular reentry circuit that uses the AV node anterogradely and a rapidly conducting accessory pathway retrogradely. Orthodromic circus movement tachycardia is the second most common mechanism of PSVT. In patients with AT, there are congenital and surgically acquired accessory pathways responsible for the increased rate of Wolff-Parkinson-White syndrome. Both types of accessory pathways can and should be treated by means of catheter ablation because atrial arrhythmia often seen in patients undergoing the Fontan operation can trigger atrioventricular re-entrant tachycardia or cause life-threatening tachycardia.

Congenital accessory pathways should be excluded carefully before surgical intervention for total cavopulmonary anastomosis in patients with AT.(1)

1. **Hager A,et al. Congenital and surgically acquired Wolff-Parkinson-White syndrome in patients with tricuspid atresia. Thorac Cardiovasc Surg. 2005 Jul;130:48-53.**

Type A Wolff-Parkinson-White. Anomalous accessory pathway is localized on left lateral wall: Between the LA and the LV Prominent anterior forces by posteroanterior activation. Why?. Answer: following d' Avila's algorithm(1) to locate accessory pathway on the basis of QRS complex polarity. It is based on QRS complex polarity analysis in 5 electrocardiographic leads, to locate the accessory bundle



Sequence of the analysis:

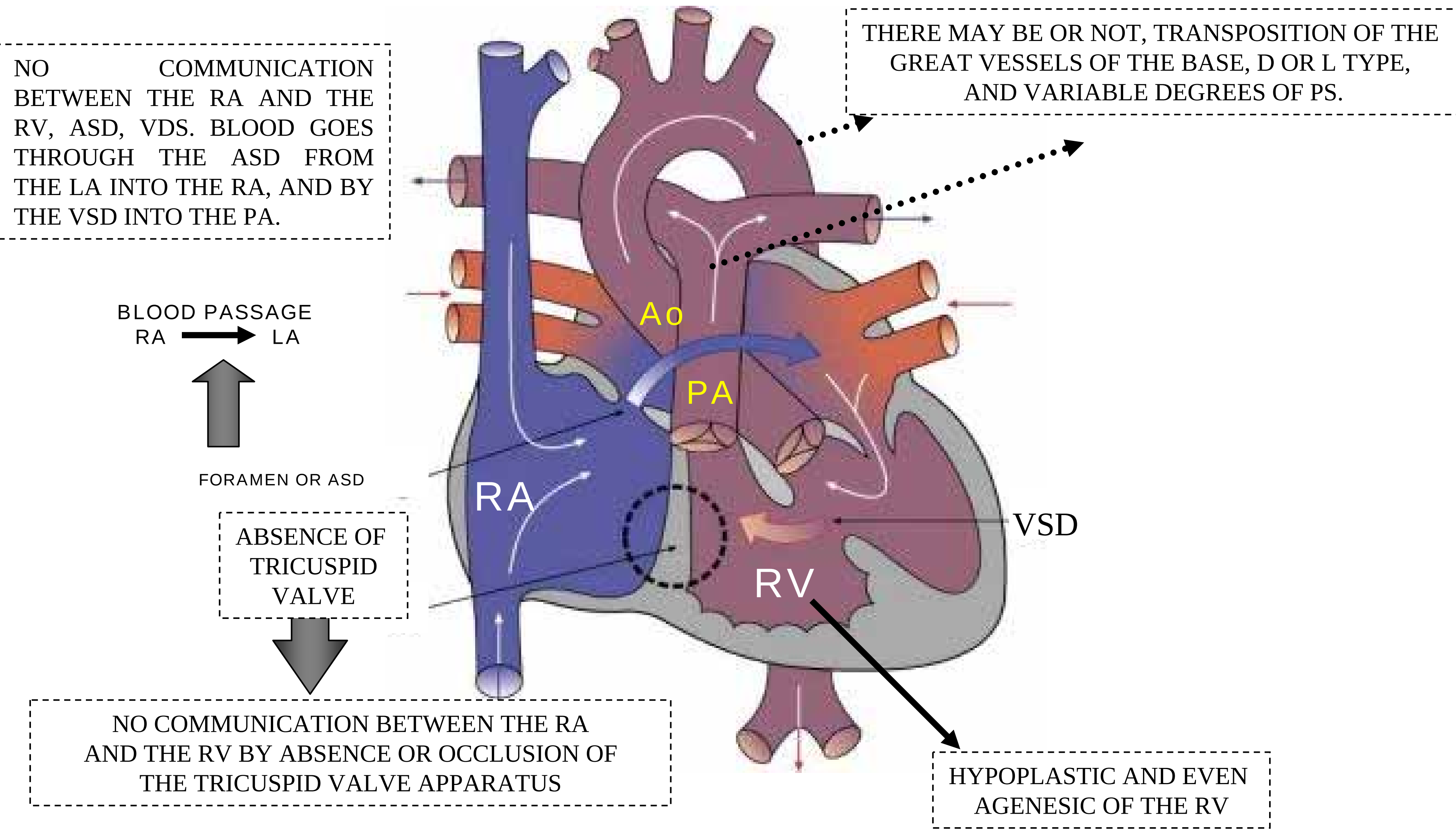
1. **STEP:** *to define QRS complex polarity in V1. Two possibilities: positive or plus-minus or negative; In the present case QRS is positive in V1 pseudo RBBBB,*

2º STEP: *in case of being positive or plus-minus in V1, the III lead should be checked next. If it is positive, this is a left lateral accessory pathway (LLAP). If it is isodiphasic plus-minus, this is a left postero-lateral pathway (PLAP). Finally, if III is negative, the AP will be left postero-septal (LPSAP).*

If polarity in V1 is negative and positive in aVF: anteroseptal (ASAP). If it is negative in aVL, it is left lateral (LLAP). If it is plus-minus in III, it is anteroseptal (ASAP).

1. d' Ávila, et al. Pacing Clin Electrophysiol 1995;18:1615-1627.

TRICUSPID ATRESIA (TA)



Outline of the situation in Tricuspid Atresia (TA): absence of tricuspid valve that predisposes RV hypoplasia and RA enlargement with short circuit from right to left between the RA and the LA, resulting in LV volume enlargement.

Tricuspid atresia

Tricuspid atresia is a form of congenital heart disease whereby there is a complete absence of the tricuspid valve. Therefore, there is an absence of right atrioventricular connection. This leads to a hypoplastic or an absence of the right ventricle.

Because of the lack of an A-V connection, an atrial septal defectl(ASD) must be present to maintain blood flow. Also, since there is a lack of a right ventricle there must be a way to pump blood into the pulmonaries arteries, and this is accomplished by a ventricular septal defect (VSD).

Blood is mixed in the left atrium. Because the only way the pulmonary circulation receives blood is through the VSD, a patent ductus arteriousus is usually also formed to increase pulmonary flow.

These babies may get some blood to their lungs by a different route. Even so, they do not get enough oxygen to their bodies, which can make them look blue (cyanotic). Also their right ventricle does not grow the way it should.

Epidemiology

Tricuspid atresia is an uncommon form of congenital heart disease that affects about 5 in every 100,000 live births.

Tricuspid atresia is the third most common form of cyanotic congenital heart disease, with a prevalence of 0.3-3.7% in patients with congenital heart disease.

Twenty percent of patients with this condition will also have other cardiovascular anomalies.

A persistent left superior vena cava anomaly is observed in 15% of patients.

Most of the associated anomalies relate to transposition of the great vessels: Groups I and II.

Frequency

The frequency of tricuspid atresia is 2.9% in autopsy series at the United States. The frequency of tricuspid atresia is 2.9% in autopsy series at the United States. Tricuspid Atresia is the 3rd commonest cyanotic congenital Heart disease.

Ethnic group

No racial predilection is apparent.

Mortality/Morbidity

Depending on the degree of obstruction and associated anomalies, TA may be lethal at birth. Without repair, the patient rarely survives to adulthood. TA has a mortality rate of 90% before the age of 10 years.(1)

Gender

Considering all forms of tricuspid atresia, no sexual predilection exists.
Males present more frequently with transposed great vessels than females (Groups II and III).

Age

The anomaly is congenital and is evident at birth.

Causes

The cause is unknown. Although specific genetic causes of the malformation remain to be determined in humans, the ZFPM2/FOG2 and HEY2 genes(2) may be involved in the process. Mice in which the FOG2 gene is knocked out are born with tricuspid atresia. The significance of this finding and its applicability in humans requires further investigation.

1. Keating P, Van der Merwe, Shipton. Tricuspid atresia--profile and outcome. Cardiovasc J S Afr. 2001 Aug-Sep;12:202-205.
2. Sarkozy A, Conti E, D'Agostino R, Digilio MC, Formigari R, Picchio F, et al ZFPM2/FOG2 and HEY2 genes analysis in nonsyndromic tricuspid atresia. Am J Med Genet A. 2005 Feb 15;133A:68-70.

Embryology

The atrioventricular valves develop shortly after the atrioventricular canal divides. The tricuspid valve leaflets have several origins. The septal leaflet of the tricuspid valve mostly develops from the inferior endocardial cushion with a small contribution from the superior cushion. The anterior and posterior tricuspid valve leaflets develop by undermining of a skirt of ventricular muscle tissue. The process of undermining extends until the atrioventricular valve junction is reached. Resorption of the muscle tissue produces normal-appearing valve leaflets and chordae tendineae.(1, 2, 3) Fusion of developing valve leaflet components results in stenosis (partial fusion) or TA (complete fusion) of the valve.(4, 5)

Whether a muscular type of TA develops or whether well-formed but fused tricuspid-valve leaflets develop depends on the stage of development when the embryologic aberration takes place. The classic muscular form of TA develops if the embryologic insult occurs early in gestation, and fused valve leaflets occur if the embryologic abnormality occurs slightly later than this in gestation. If the valve fusion is incomplete, stenosis of the tricuspid valve develops. Therefore, the fact that isolated congenital tricuspid stenosis belongs to the group of TA defects and that their embryologic developments are similar is no surprise. Thus, the tricuspid valve stenosis, TA with well formed but fused valve leaflets, and the muscular type of TA represent a spectrum of morphologic abnormalities.(4 5). The pathologic, clinical, and ECG features of tricuspid stenosis and TA are similar.(6)

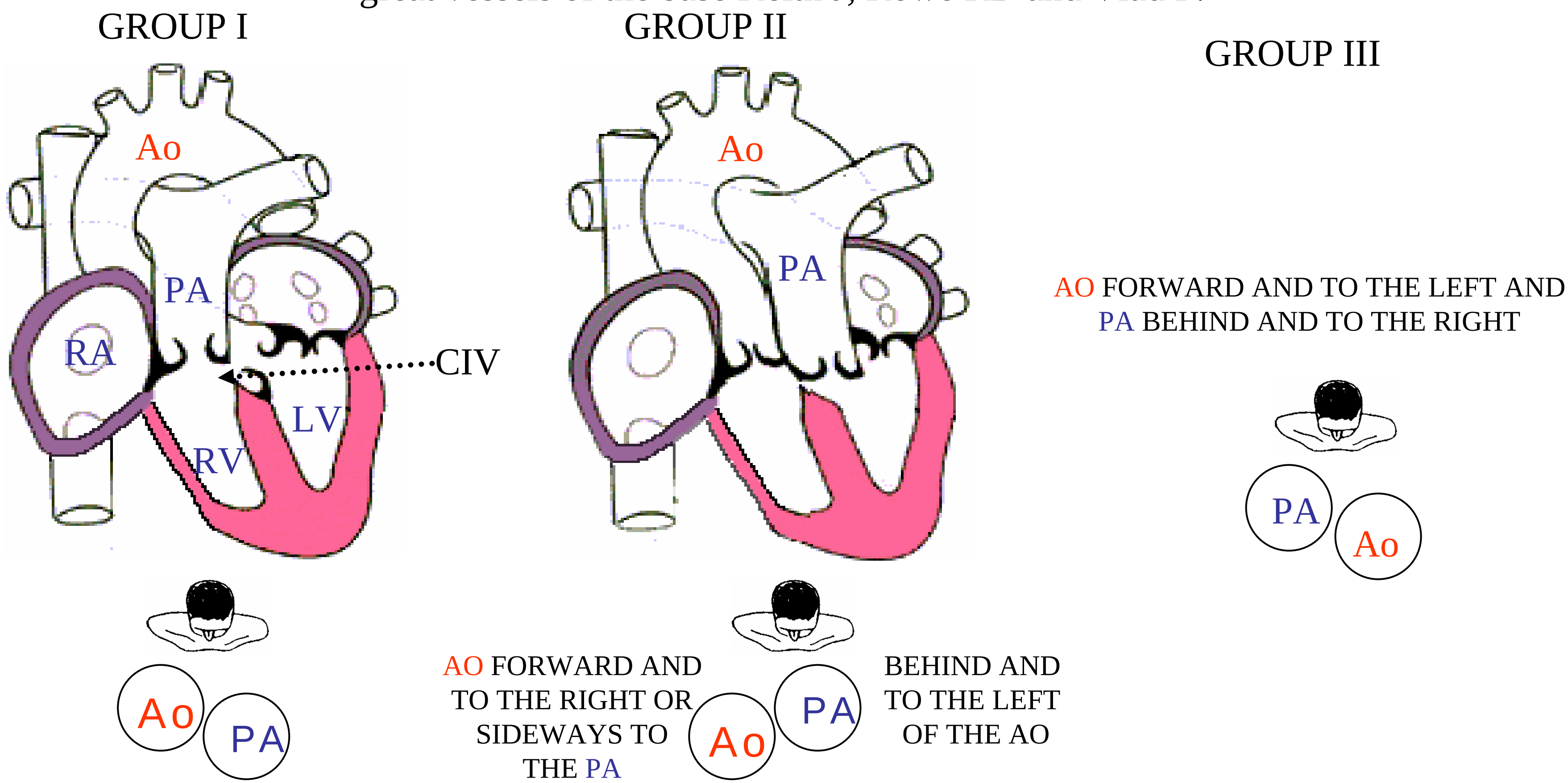
1. *Ando M, Santomi G, Takao A. Atresia of tricuspid and mitral orifice: anatomic spectrum and morphogenetic hypothesis. In: Van Praagh R, Takao A, eds. Etiology and Morphogenesis of Congenital Heart Disease. NY: Futura: Mount Kisco; 1980: 421-87.*
2. *Van Mierop LH, Gessner IH. Pathogenetic mechanisms in congenital cardiovascular malformations. Prog Cardiovasc Dis. Jul-Aug 1972;15:67-85.*
3. *Wilson AD, Rao PS. Embryology. In: Kambam J, ed. Cardiac Anesthesia for Infants and Children. St Louis, Mo: Mosby; 1994:3-9.*
4. *Rao PS. Tricuspid atresia. In: Long WA, ed. Fetal and Neonatal Cardiology. Philadelphia, Pa: WB Saunders; 1990:525-40.*
5. *Rao PS, Alpert BS, Covitz W. Left ventricular function in tricuspid atresia. In: Rao PS, ed. Tricuspid Atresia. 2nd ed. Mount Kisco, NY: Futura; 1992:247-59.*
6. *Keefe JF, Wolk MJ, Levine HJ. Isolated tricuspid valvular stenosis. Am J Cardiol. Feb 1970;25:252-7*

Subtypes

TA is classified according to the morphology of the valve,(1) the radiographic appearance of pulmonary vascular markings,(2; 3) and the associated cardiac defects.(4, 5, 6, 7, 8)
Van Praagh et al (9) initially proposed a classification based on the morphology of the atretic tricuspid valve. He and others later modified and expanded the classification, as described in TA.(10)
Astley et al. (2) proposed a classification based on pulmonary vascular markings on a chest radiograph: Group A are cases with decreased pulmonary vascular markings, and group B are those with increased pulmonary vascular markings. (2) Dick et al (3) added a third group, group C, to describe cases with a transition from increased to decreased pulmonary vascular markings. This type of classification has some clinical value, although a more precise definition than these can often be made by using noninvasive 2-dimensional (2D) and Doppler echocardiography.
In 1906, Kuhne (4) first proposed a classification based on great-artery relationships, which Edwards and Burchell expanded in 1949.(10)Keith, Rowe, and Vlad popularized the following classification in 1967.(11)

1. Van Praagh R. Discussion after paper by Vlad P: Pulmonary atresia with intact ventricular septum. In: Barrett-Boyes BG, Neutze JM, Harris EA, eds. *Heart Disease in Infancy: Diagnosis and Surgical Treatment*. London: Churchill Livingstone; 1973:236.
2. Astley R, Oldham JS, Parsons C. Congenital tricuspid atresia. *Br Heart J*. Jul 1953;15(3):287-297.
3. Dick M, Fyler DC, Nadas AS. Tricuspid atresia: clinical course in 101 patients. *Am J Cardiol*. Sep 1975;36(3):327-37.
4. Kuhne M. Uber zwei falle kongenitaler atreside des ostium venosum dextrum. *Jahrb Kinderh*. 1906;63:235.
5. Edwards JE, Burchell HB. Congenital tricuspid atresia: a classification. *Med Clin North Am*. 1949;33:1117.
6. Keith J, Rowe RD, Vlad P. Heart Disease in Infancy and Childhood. In: *Tricuspid Atresia*. 2nd ed. New York: Macmillian; 1967:434, 664.
7. Vlad P. Tricuspid atresia. In: Keith JD, Rowe RD, Vlad P, eds. *Heart Disease in Infancy and Childhood*. 3rd ed. New York: Macmillian; 1977:518-41.
8. Rao PS. A unified classification for tricuspid atresia. *Am Heart J*. Jun 1980;99(6):799-804
9. Van Praagh R, Ando M, Dungan WT. Anatomic types of tricuspid atresia: clinical and developmental implications [abstract]. *Circulation*. 1971;44:115.
10. Edwards JE, Burchell HB. Congenital tricuspid atresia: a classification. *Med Clin North Am*. 1949;33:1117.
11. Keith J, Rowe RD, Vlad P. Heart Disease in Infancy and Childhood. In: *Tricuspid Atresia*. 2nd ed. New York: Macmillian; 1967:434, 664.

Classification of the three TA groups, taking into account the presence or not of D or L-transposition of the great vessels of the base Keith J, Rowe RD and Vlad P.



By the presence or absence of transposition and its characteristics, Keith, Rowe and Vlad grouped TA in three groups:

Group I: TA with TA normally related great arteries no transposition of the great vessels of the base: 70%;

Group II: TA with D-transposition: 27%;

Group III: TA with L-transposition: 3%.

1. Keith J, Rowe RD, Vlad P. Heart Disease in Infancy and Childhood. In: *Tricuspid Atresia*. 2nd ed. New York: Macmillian; 1967:434, 664.

The three antomical groups and their subvarieties without transposition of the great vessels, with D-transposition or L- transposition: pulmonary atresia, pulmonary stenosis and without pulmonary stenosis

Group I or type 1: TA normally related great arteries

Ia: Pulmonary Atresia (no VSD with PDA);

Ib: Pulmonary Stenosis (with small Ventricular Septal Defect (VSD);

Ic: Without pulmonary stenosis (with extensive VSD).

Group II: TA with D-transposition

IIa: Pulmonary Atresia, extensive VSD and PDA;

IIb: Pulmonary Stenosis and extensive VSD;

IIc: Normal PA and extensive VSD.

Group III: TA with L- transposition

IIIa: With Subpulmonary Stenosis;

IIIb: With Sub-aortic Stenosis.

Patients with increased pulmonary blood flow have better prognosis than those with decreased pulmonary blood flow.

1. Keith J, Rowe RD, Vlad P. Heart Disease in Infancy and Childhood. In: *Tricuspid Atresia*. 2nd ed. New York: Macmillian; 1967:434, 664.
2. Rao PS. A unified classification for tricuspid atresia. Am Heart J. 1980 Jun;99:799-804.

Other classifications are reviewed in the American Heart Journal(1) and TA.(2) Although these classifications are generally good, their exclusion of some variations in great-artery relationships and the lack of consistency in subgroups are problematic.

Mitchell et al. (3) propose the following new, comprehensive-yet-unified classification: The principle grouping continues to be based on the following interrelationships of the great arteries:

Type I - Normally related great arteries

Type II - D-Transposition of the great arteries

Type III - Great artery positional abnormalities other than D-transposition of the great arteries

Subtype 1 - L-Transposition of the great arteries

Subtype 2 - Double outlet right ventricle

Subtype 3 - Double outlet left ventricle

Subtype 4 - D-malposition of the great arteries (anatomically corrected malposition)

Subtype 5 - L-malposition of the great arteries (anatomically corrected malposition)

Type IV - Persistent truncus arteriosus

All types and subtypes are subdivided into the following subgroups:

Subgroup a - Pulmonary atresia

S ubgroup b - Pulmonary stenosis or hypoplasia

Subgroup c - No pulmonary stenosis (normal pulmonary arteries)

This unified classification includes all the previously described abnormalities in the positions of the great arteries and can be further expanded if new variations are revealed. This classification maintains uniformity of the subgroups and preserves the basic principles of classification that Kuhne, Edwards and Burchell, and Keith, Rowe, and Vlad devised.

1. *Rao PS. A unified classification for tricuspid atresia. Am Heart J. Jun 1980;99:799-804.*
2. *Rao PS, Covitz W, Chopra PS. Principles of palliative management of patients with tricuspid atresia. In: Rao PS, ed. Tricuspid Atresia. 2nd ed. Mount Kisco, NY: Futura; 1992:297-320.*
3. *Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Incidence and natural history. Circulation. Mar 1971;43:323-332.*

Associated cardiac defects in tricuspid atresia (1)

D-Transposition of the great arteries
L-Transposition of the great arteries
Double outlet right ventricle
Double outlet left ventricle
Other malpositions of the great arteries
Truncus arteriosus

Defects that may need attention before or during palliative or total surgical correction

Absent pulmonary valve
Aneurysm of the atrial septum
Aortopulmonary fistula
Coarctation of the aorta
Common atrium
Cor triatriatum dexter
Coronary sinus septal defect
Double aortic arch
Double-outlet left atrium
Hemitruncus
Hypoplastic ascending aorta and/or aortic atresia
Ostium primum ASD
Parchment right ventricle
Patent ductus arteriosus
Anomalous origin of the coronary arteries from the pulmonary artery
Anomalous origin of the left subclavian artery
Anomalous origin of the right subclavian artery
Persistent left superior vena cava
Right aortic arch
Subaortic stenosis
Total anomalous pulmonary venous connection
Tubular hypoplasia of the aortic arch
Valvar aortic stenosis

Other associated defects are as follows:

Juxtaposition of the atrial appendages
Anomalous entry of coronary sinus into the left atrium

1. Rao PS, Covitz W, Chopra PS. Principles of palliative management of patients with tricuspid atresia. In: Rao PS, ed. *Tricuspid Atresia*. 2nd ed. Mount Kisco, NY: Futura; 1992:297-320.

History

Tricuspid atresia is usually detected in infancy because of presenting cyanosis, congestive heart failure, and growth retardation. Parents provide a history of poor skin coloration (ranging from pallor to frank cyanosis), inability to complete a feeding session, frequent pauses during feeding, and/or frank anorexia. As a result, the infant demonstrates poor growth patterns. Respiratory difficulties are often reported as nasal flaring or muscle retractions..) Bacterial endocarditis and brain abscess are common findings in patients with tricuspid atresia and should be considered in children with headaches, seizures, or neurologic deficits.

Physical

On inspection, cianosis is the most common clinical feature of this lesion. The degree of cyanosis depends on the degree of pulmonary blood flow. Infants with associated diminished pulmonary blood flow or infants who depend on a patent ductus arteriosus manifest pronounced cyanosis that worsens as the ductus begins to close. Patients with relatively normal or increased pulmonary blood flow manifest little cyanosis but more pronounced congestive heart failure. Digital clubbing is common in infants older than 3 months. Jugular venous pulsations and distention are common.

The peripheral pulses and pulse volume may be decreased, normal, or increased. The left ventricular impulse is prominent because of volume overload. The apical impulse is hyperdynamic, with displacement to the left of the midclavicular line. A thrill may be felt at the left sternal border in patients with a restrictive ventricular septal defect or pulmonary valve stenosis. The liver may be large and pulsatile.

A single first heart sound that may be increased in intensity is usually present.

The second heart sound may be single or normally split. The intensity of this sound varies, depending on associated transposition of the great vessels. In normally related great vessels, the second heart sound may be of normal intensity. In transposed great vessels, the second sound is diminished.

Cardiac murmurs are present in 80% of patients with tricuspid atresia: A holosystolic murmur that may have a crescendo and decrescendo quality is present, signifying blood flow through the ventricular septal defect.

A continuous murmur may be present. Systemic-to-pulmonary arterial collaterals or arterial-to-pulmonary arterial anastomoses surgically created to improve pulmonary blood flow may cause this finding.

A murmur of mitral insufficiency may also be present. Holosystolic murmur due to the VSD.

Electrocardiogram in Tricuspid Atresia

P wave

Right atrial enlargement: *Right atrial overload is manifest as tall with abnormally high voltage P waves in lead II. It is frequent in infants. Its height does not correlate well with atrial pressure or with the size of the atrial septal defect. There are descriptions of very high P waves(1)*
The biphasic P wave in V1 with sharp atrial intrinsicoid deflection (the sharp downward deflection from the peak of the P wave to the trough of the P wave) is a pseudo left atrial overload pattern, seen in right atrial overload. In true left atrial overload the atrial intrinscoid deflection is more slanting so that the negative component of the P wave is almost U shaped rather than the V shape in this case.

Biatrial enlargement: *tall and notched P waves was observed in approximately 80% of cases. (2)*

1. Reddy SC, Zuberbuhler JR. Images in cardiovascular medicine. Himalayan P-waves in a patient with tricuspid atresia. Circulation. 2003 Jan 28;107(3):498.
2. Gamboa R, Gersony WM, Nadas AS. The electrocardiogram in tricuspid atresia and pulmonary atresia with intact ventricular septum. Circulation. 1966 Jul;34:24-37.
3. Dick M, Fyler DC, Nadas AS. Tricuspid atresia: clinical course in 101 patients. Am J Cardiol. 1975 Sep;36:327-337.

PR interval

Relatively short PR interval is observed un 50% of cases in AT. Additionally, slurring of the initial QRS complex is observed suggesting preexcitation. Zeller et al (1) conducted a retrospective study to determine the frequency of this ECG pattern and whether or not this represented the presence of a true atrioventricular bypass tract. Three pediatric cardiologists reviewed the surface ECGs of 183 consecutive AT patients. The patients' ages ranged from 4 months to 21 years.
The criteria for preexcitation included:

1) *PR segment <100ms*
2) *A QRS complex ≥100ms and*
3) *Slurring of the upstroke of the QRS complex ("delta wave").*

Of the 183 patients, 22 (12%) had PR segments < 100ms, 9 of whom fulfilled the criteria for preexcitation. Five of these had a history of supraventricular tachycardia, and 4 of the 5 had undergone invasive electrophysiologic studies: 2 had enhanced atrioventricular-nodal conduction and 1 had normal atrioventricular-nodal conduction; only 1 had an accessory pathway.
These results indicate that, although many patients with TA meet the surface ECG criteria for preexcitation, many of these patients may not have an atrioventricular bypass tract; this state might be termed "pseudo-preexcitation“. In these instances, invasive studies probably would not be necessary; regrettably, it may be difficult to distinguish between the presence and the absence of preexcitation in such patients without invasive electrophysiologic studies.

1. **Zellers TM, Porter CB, Driscoll DJ. Pseudo-preexcitation in tricuspid atresia. Tex Heart Inst J. 1991;18:124-126.**

QRS axis on frontal plane

QRS axis between 0º and -90º with counter clock wise rotation of QRS loop in frontal plane is observed in ≈85% of cases. In general left axis deviation is common anatomic type(Ib) and when D-transposition is present

Guller et al(1) of two cases of AT in whom sections of the AV node conducting system were studied ,it was found that the left bundle branch originated very early from the common bundle and that the right bundle branch was markedly elongated. The unusual early origin of the left bundle branch could explain an abnormal sequence of depolarization leading to left axis deviation and a counterclockwise QRS loop in the frontal plane. Additionally, this early left bundle branch block origin is responsible by short PR interval tendency observed(1)

A QRS axis between 0º and +90º is observed in 7% of cases. It is more frequent in cases with transposition and increased pulmonary flow.

In 4% of case right axis deviation is present. It is indicative of large right ventricle and large pulmonary flow or pulmonary hipertension.

1. **Guller B, Dushane JW, Tifus JLThe atrioventricular conduction system in two cases of tricuspid atresia. Circulation. 1969 Aug;40:217-226.**

Precordial leads

Characteristic an adult pattern of QRS progression over the precordial leads (V1 through V6).
Left ventricular dominance. LVH/LVE Eventual positive Sokolow-Lyon criteria. S wave of V1 + R of V5 ≥ 35 mm or 3.5 mV in adults older than 30, > 40 mm or 4.0 mV between 20 and 30 years (Sokolow-Rapaport), > 60 mm between 16 and 20 years and > than 65 mm between 11 and 16 years. Sensitivity: 25%. Specificity: 95%.
Inverted T waves in left leads(Strain pattern) is observed in ≈45% of cases.
QRS loop of VCG in HP has counterclockwise wise rotation, leftward and posteriorly. This pattern of QRS is characterized by absent right ventricular forces and well-developed left ventricular forces consistent with LVH.
The presence of righward terminal forces and maximal spatial vector (LMSV) directed to front is indicative of relatively large RV and increased pulmonary flow (only 1% of cases).

Wolff-Parkinson-White syndrome

In patients with AT, there are congenital and surgically acquired accessory pathways responsible for the increased rate of Wolff-Parkinson-White syndrome. Both types of accessory pathways can and should be treated by means of catheter ablation because atrial arrhythmia often seen in patients undergoing the Fontan operation can trigger atrioventricular re-entrant tachycardia or cause life-threatening tachycardia. Congenital accessory pathways should be excluded carefully before surgical intervention for total cavopulmonary anastomosis in patients with AT.(1)

1. **Hager A, Zrenner B, Brodherr-Heberlein S, Steinbauer-Rosenthal I, Schreieck J, Hess J.J Congenital and surgically acquired Wolff-Parkinson-White syndrome in patients with tricuspid atresia. Thorac Cardiovasc Surg. 2005 Jul;130:48-53.**

The Fontan-type procedure and its effect on ECG

This procedure has undergone 2 major modifications, including intra-atrial baffling and extracardiac conduit. To clarify the effect of these modifications on arrhythmia propensity, Koh et al (1) analyzed chronologic changes in P-wave characteristics after atriopulmonary connection, intra-atrial baffling, or extracardiac conduit. After intra-atrial baffling, patients increasingly had prolonged P-wave duration and larger dispersion associated with sinus node dysfunction (SND), suggesting a propensity to arrhythmia, although less progressive than seen in those undergoing atriopulmonary connection. In contrast, despite an equal prevalence of SND after extracardiac conduit, the lack of important changes in P-wave characteristics over time suggests that the extracardiac conduit procedure is the preferred option for optimal rhythm prognosis. Early atrial dysrhythmias after the Fontan operation are related to preoperative hemodynamics. Early supraventricular tachycardia/SND and the atriopulmonary type of Fontan connection are significant risk factors for late atrial dysrhythmias.(2)

Patients with atrial tachyarrhythmias late after Fontan operation have longer P-wave duration and P-wave dispersion and larger right atrial dimension than those without the arrhythmias; these abnormalities are interrelated. This observation represents an atrial mechano-electrical remodeling phenomenon in parallel to an increase in arrhythmia propensity in this vulnerable population and warrants further investigation.(3)

1. ***Koh M, Uemura H, Kada A, Kagisaki K, Hagino I, Yagihara T.Chronologic changes in P-wave characteristics after the Fontan procedure: the effect of surgical modification. J Thorac Cardiovasc Surg. 2010 Jul;140:137-143.***
2. ***Paul T, Ziemer G, Luhmer I, Hecker H, Kallfelz HC. Atrial arrhythmias after modified Fontane operation: effect of preoperative hemodynamics and the kind of operation (atriopulmonary vs. total cavopulmonary Z Kardiol. 1993 Jun;82:368-375.***
3. ***Wong T, Davlouros PA, Li W, Millington-Sanders C, Francis DP, Gatzoulis MA.. Mechano-electrical interaction late after Fontan operation: relation between P-wave duration and dispersion, right atrial size, and atrial arrhythmias. Circulation. 2004 May 18;109:2319-2325.***

ELECTROCARDIOGRAPHIC CLUES

P wave of right atrial enlargement (RAE): P "tricuspidale" of Gamboa. LV enlargement of volumetric type (diastolic LVE) with possible adult progression in precordial leads.

Pattern of left anterior fascicular block of the His bundle (LAFB): extreme shift in the left superior quadrant of AQRS and counterclockwise rotation of the QRS loop in the FP. From 120 children with TA the ECG with left axis axis deviation was observed in 94%, RAE in 58%, LVE in 96% and LAE: 47.5%.(1)

Association of:

Right atrial enlargement/ hypertrophy/overloading (RAE or BAE)

+

Left ventricular hypertrophy/enlargement (LVH/LVE) or excessive LV dominance for age

(The right ventricle is ussually hypoplastic, but if there is a large VSD that is present and the presence or absence of pulmonary stenosis)

+

Extreme shift fo QRS in the left superior quadrant and counterclockwise rotation in the FP.
Left anterior fascicular block ECG-pattern

+

Cyanotic baby

⇩

Highly suggestive of trcuspid atresia

1. **Rosado-Buzzo AA, Santamaría-Díaz H, Gómez-Gómez M, Alva-Espinosa C, Maulen-Radovan X, Palacios-Macedo X. Tricuspid atresia. Clinical course in 120 childrenArch Inst Cardiol Mex. 1987 Sep-Oct;57:375-381.**

Vectorcardiographic features

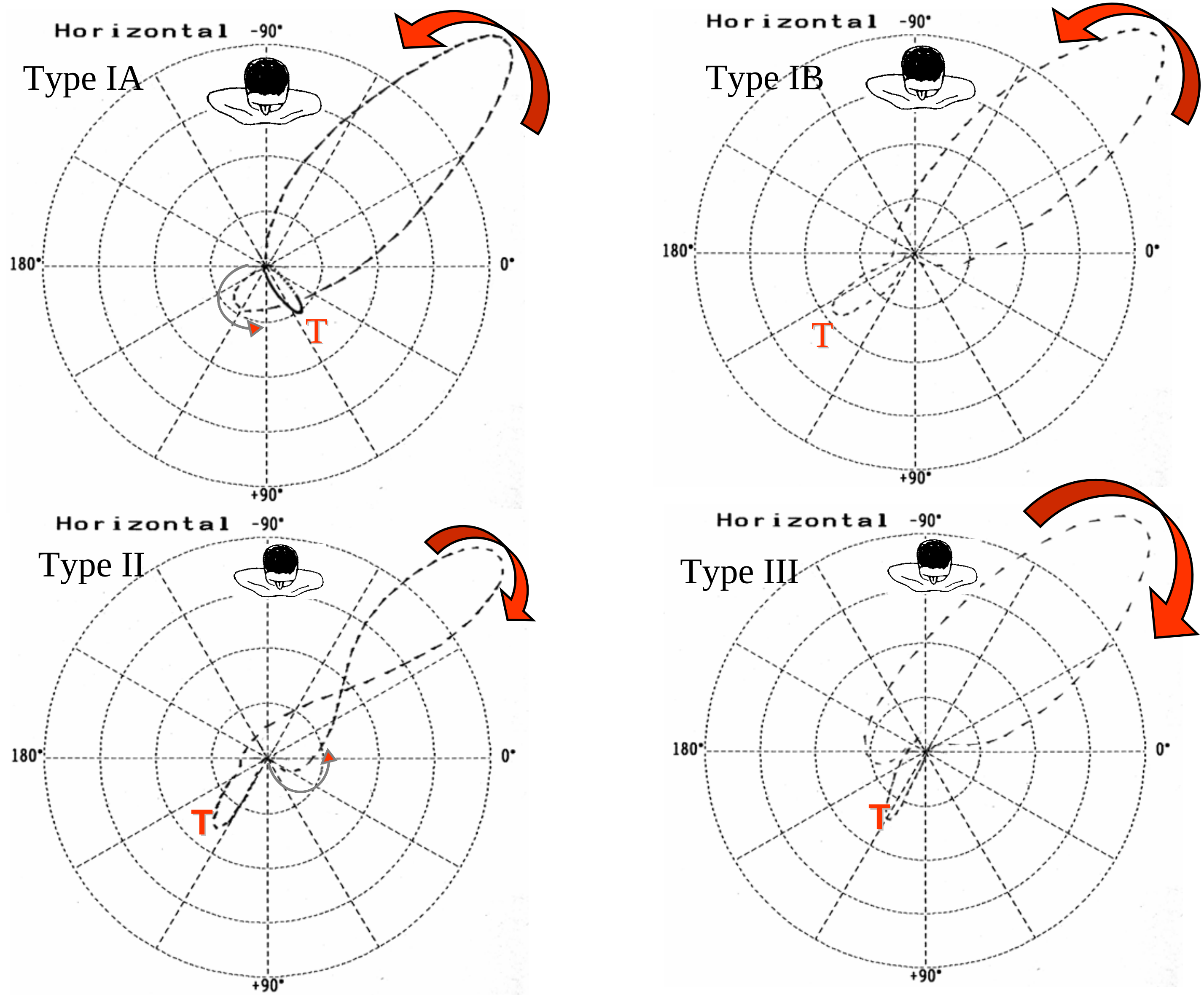
Davachi et al(1) suggested that wide loop in all planes are associated with increased pulmonary flow and narrow loops indicate reduced pulmonary flow.
The maximum spatial voltage to the left (LMSV) is increase in magnitude in all of cases(>2mV) and usually directed to back. When LMSV has anterior orientation is indicative of large pulmonary blood flow
In both groups with and without transposition al all ages a slight prolongation of the QRS duration above normal was found. The highest value in adolescent is 107ms.
The presence of right terminal forces indicate the presence of a moderate sized right ventricle and probably a large pulmonary blood flow.

Horizontal plane

P max. ant. forces≥ 0.07 mV. Maximal vector of P may exceed > 0.1 mV and CCW rotation.
QRS loop more oriented to back that normal loop
QRS loop rotation usually CCW rotation, occasionally figure in eight or even CW rotation
Frequent marked decrease of early anterior leftward forces
Sometimes had the appearance of an anterolateral myocardial infarction or Left bundle branch block
When pulmonary blood flow is augmented with large VSD and larger than average RV cavities the LMSV is dislocated anteriorly and the terminal forces located on right posterior quadrant. (3;4)

1. Davachi F, Lucas RV Jr, Moller JH. The electrocardiogram and vectorcardiogram in tricuspid atresia. Correlation with pathologic anatomy. Am J Cardiol. 1970 Jan;25:18-27.
2. Malo Concepcion P, Villa JG, Calabro R, Moreno Granado F. The electrocardiogram and the vectorcardiogram in tricuspid atresia Rev Esp Cardiol.1975 Jul-Aug;28:323-340.
3. Ellison RC ad Restieaux NJ Vectorcardigraphy in Congenital Heart Disease. A Method for Estimating Severity 1972 W.B. Saunders Company Philadelphia. London. Toronto. Chap 15 Tricuspid Atresia pp 170-175.

VECTOCARDIOGRAPHIC TYPES OF LVE/LVH IN THE HORIZONTAL PLANE



Vectocardiographic types of LVE in the horizontal plane (IA, IB, II and III).

Frontal plane

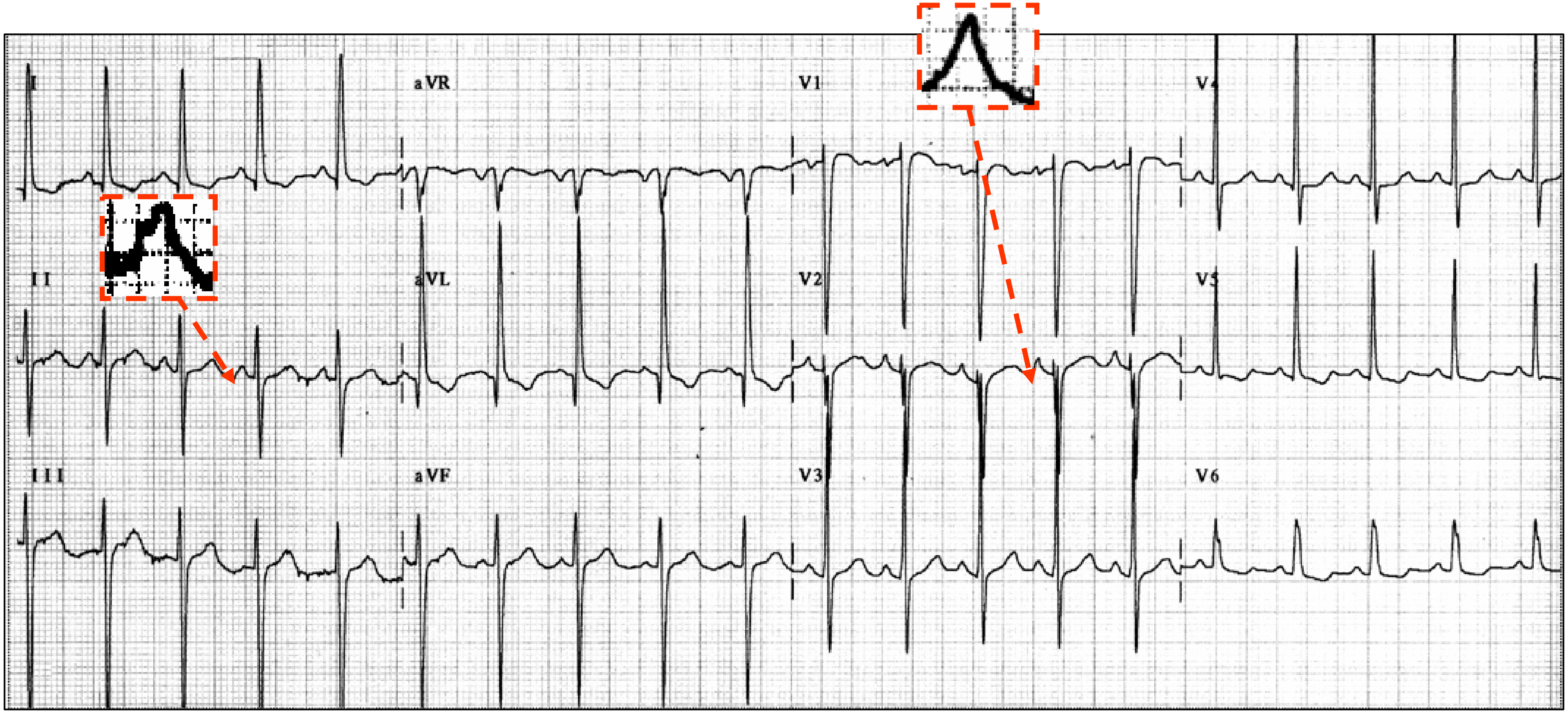
P loop is more vertical than normal, The maximal vector of P exceeds > 0.2 MV and CCW rotation
Left QRS loop axis deviation. RAE is frequent in infant and biatrial enlargement (BAE) in older patients.
Counterclockwise rotation and superiorly displaced similar to endocardial cushion defects
Rarely, ≈ 7% of cases QRS axis is between 0° to +90° This axis is observed in cases with D-transposition of great arteries association.

Right sagittal plane

Rotation is predominantly CW rotation and oriented superiorly and posteriorly
Maximal vector ≥ 1.6 mV

EXAMPLES

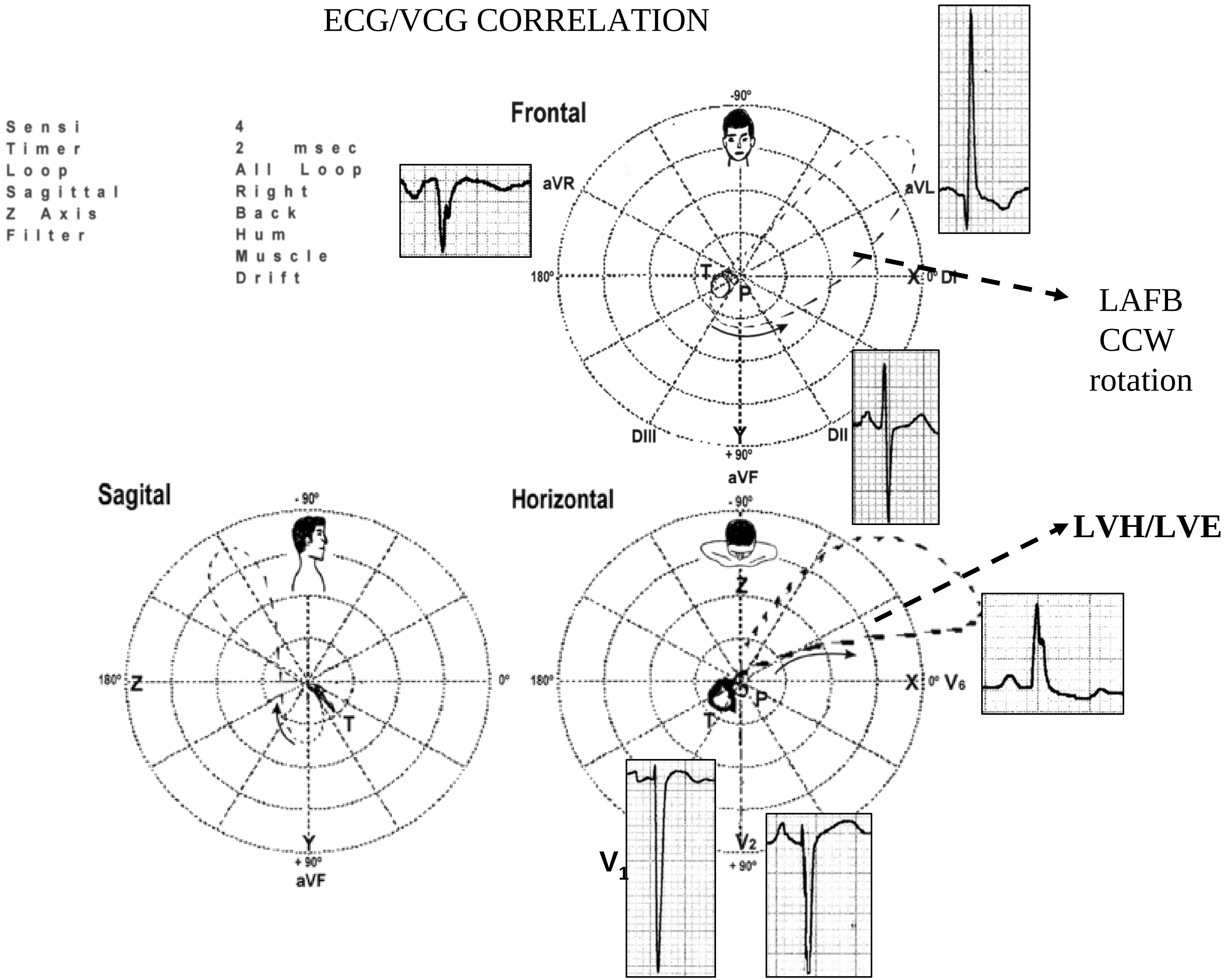
Name: LBR**; Age:** 4 y.o. **Sex:** F**. Race:** W. **Date:** 12/13/2001 **Weight:** 16 Kg. **Height:** 1.10 m
Biotype: N **Medication in use:** no use of medication.



RAE: visible in V_2 and with notch in the ascending limb of P wave. LVE/LVH: deep S in V_1 and R of increased voltage in V5. In V6 is similar to Incomplete LBBB. LAFB: SÂQRS with extreme shift in the left superior quadrant and counterclockwise rotation in the FP. qR in I and aVL. rS in inferior leads.

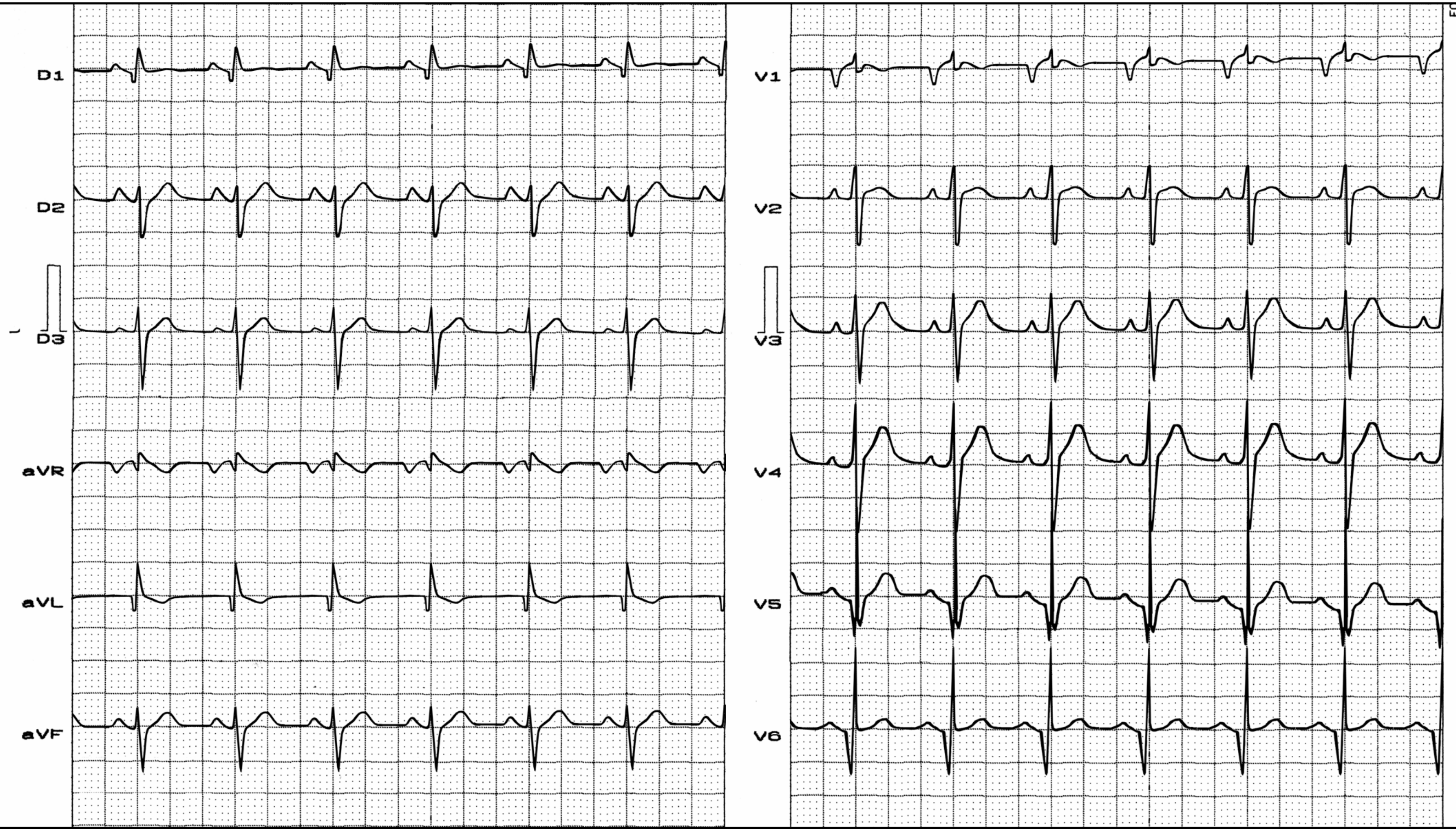
ECG/VCG of TA in a four-year-old child: RAE + LVE/LVH + LAFB.

ECG/VCG CORRELATION



ECG/VCG of tricuspid atresia in a four-year-old child: RAE + LVH/LVE + LAFB.

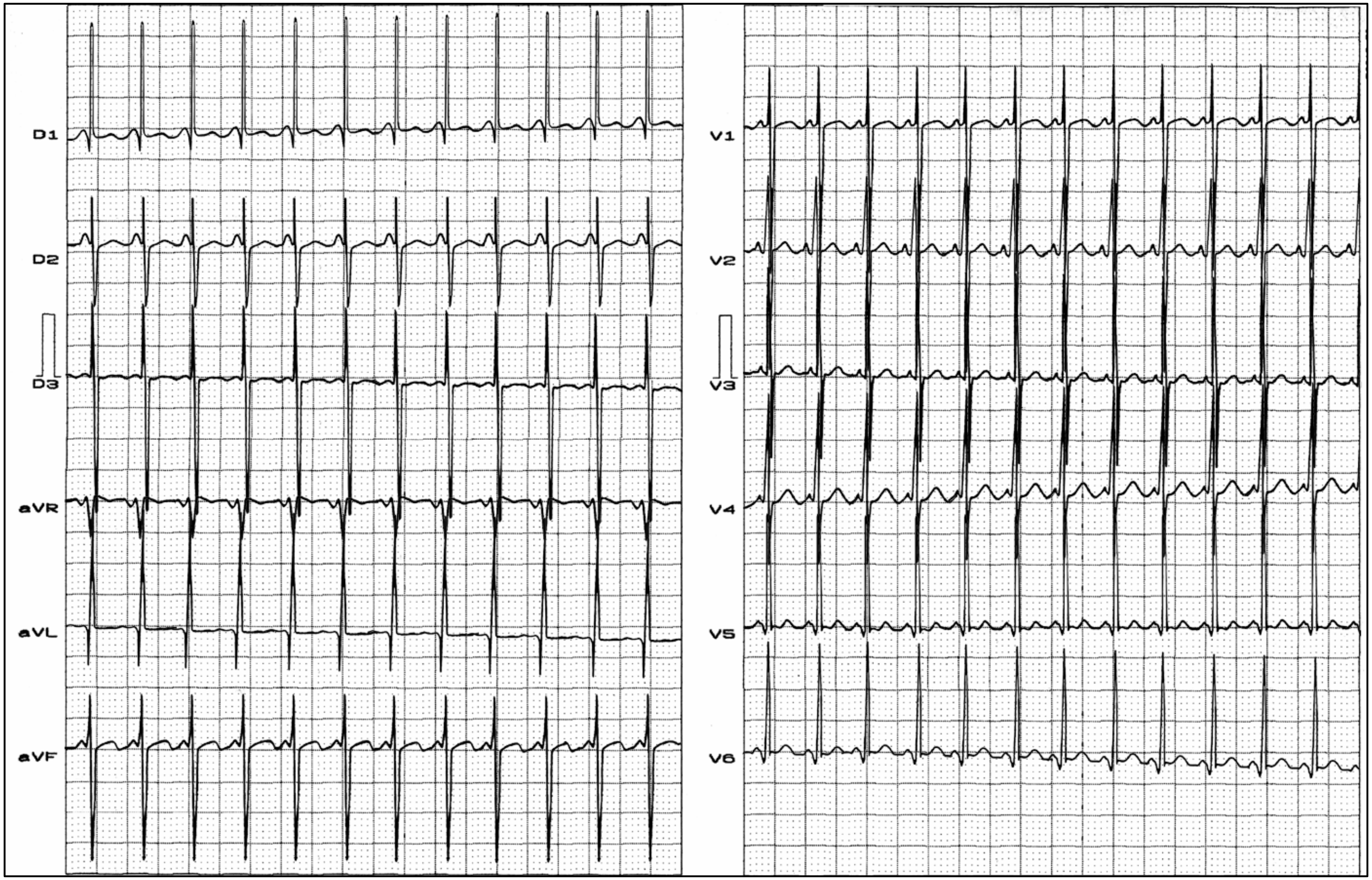
TRICUSPID ATRESIA



P WAVE: "tricuspidale of Gamboa" SAP +60º, great voltage in II and V_2 - V_4. Deep negative component in V_1. QRS: axis of QRS with extreme shift in the left superior quadrant -65º: LAFB + LVH of the diastolic type: deep Q waves in V5-V6, I and aVL.

ECG of TA with P wave of "Gamboa" + LAFB + diastolic-type LVH/LVE.

Name: DS.; **Age**: 20 days**.; Gender**: M.**; Race**: Asian. **Weight:** 2,600 gr. **Height**: 43 cm. **Date**:1/4/2000.



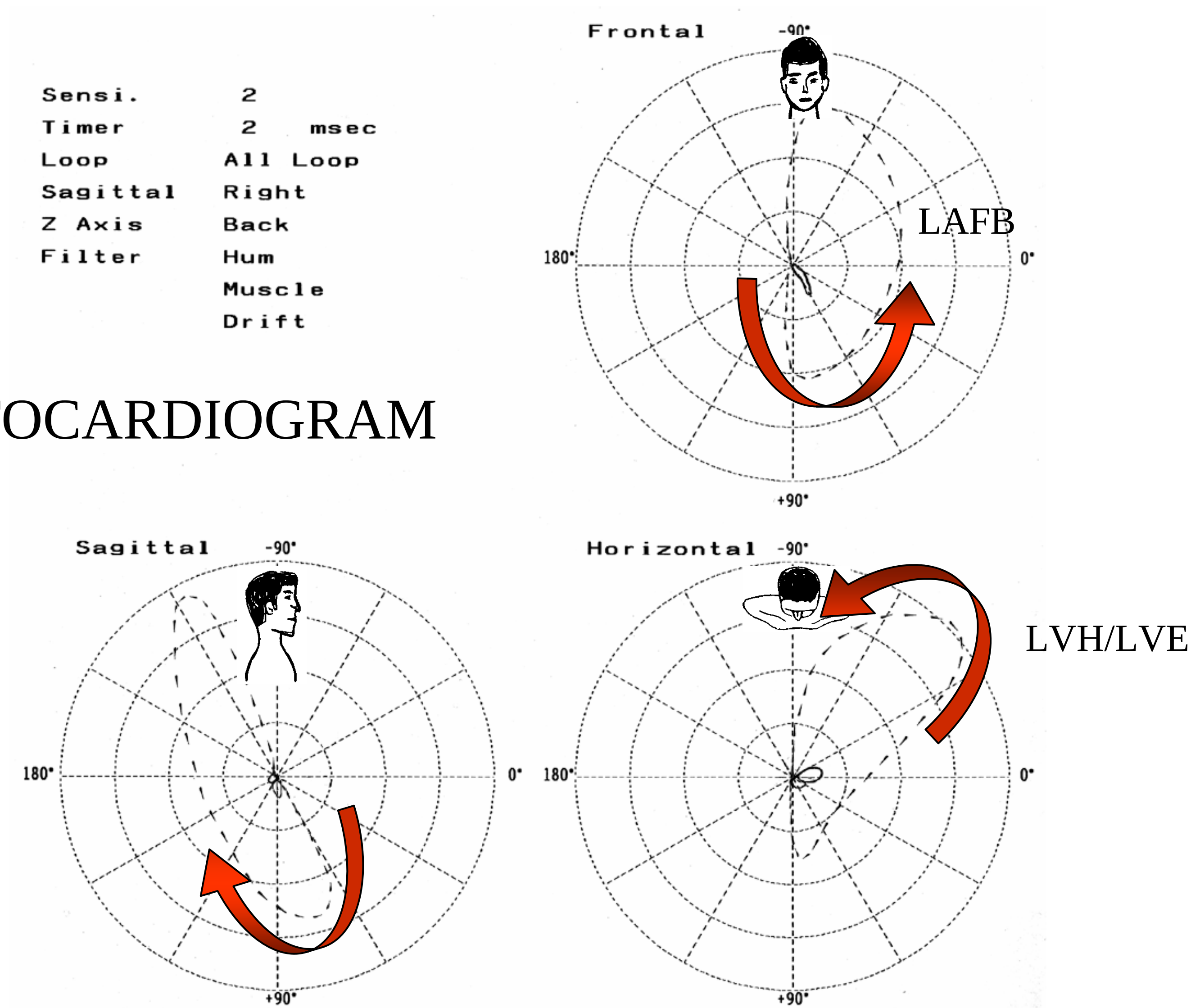
CLINICAL DIAGNOSIS: tricuspid atresia (TA). Cyanotic newborn baby.

ECG DIAGNOSIS: SR; HR: 187 bpm; P wave: voltage 2 mm and 60 ms duration; PR: tendency to be short, 70 ms (in normal newborn babies, the minimal value is 80 ms) as a consequence of congenitally short AV node and early onset of the left branch of the His bundle; QRS: SAQRS with extreme shift in the left superior quadrant; counterclockwise rotation (qR in DI and aVL). SIII > SII: LAFB: it is present in 80% of the cases of TA. There are criteria of LVE and adult progression of QRS in precordial leads.

ECG/VCG of Tricuspid Atresina in a newborn baby, cyanotic, 20 days of life. LAFB + LVE/LVH + adult progression in precordial leads. typical LAFB in the FP and typical diastolic LVE/LVH in the HP.

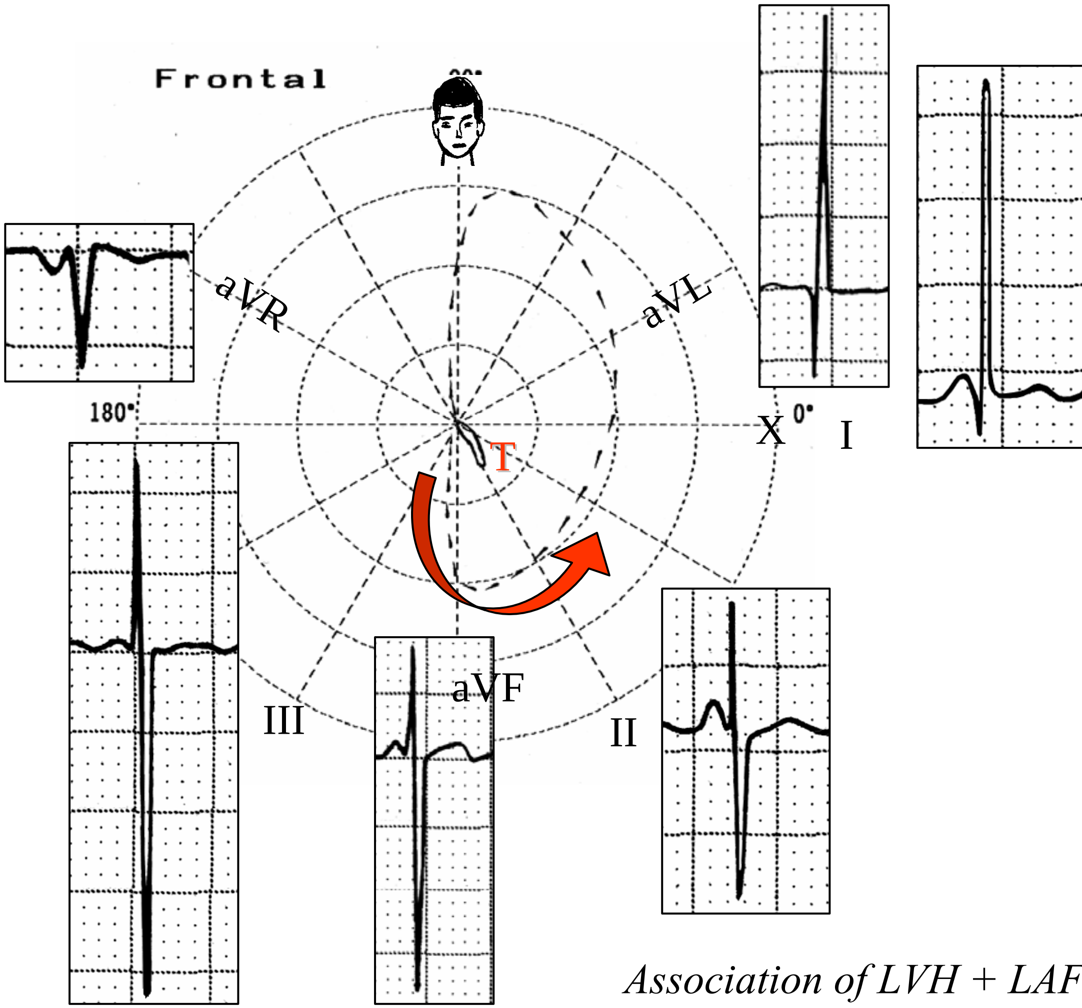
Name: DS.; **Age**: 20 days.; **Sex**: M.; **Race**: Y.; **Weight:** 2,600 gr. **Height**: 43 cm.; **Date**: 1/4/2000.

VECTOCARDIOGRAM



ECG/VCG of TA in a newborn baby, cyanotic, 20 days of life. LAFB + LVE/LVH + adult progression in precordial leads. Typical LAFB in the FP and typical diastolic LVE in the HP.

ECG/VCG CORRELATION FRONTAL PLANE



Extreme shift of QRS axis in the left superior quadrant and counterclockwise rotation in QRS loop in the FP.

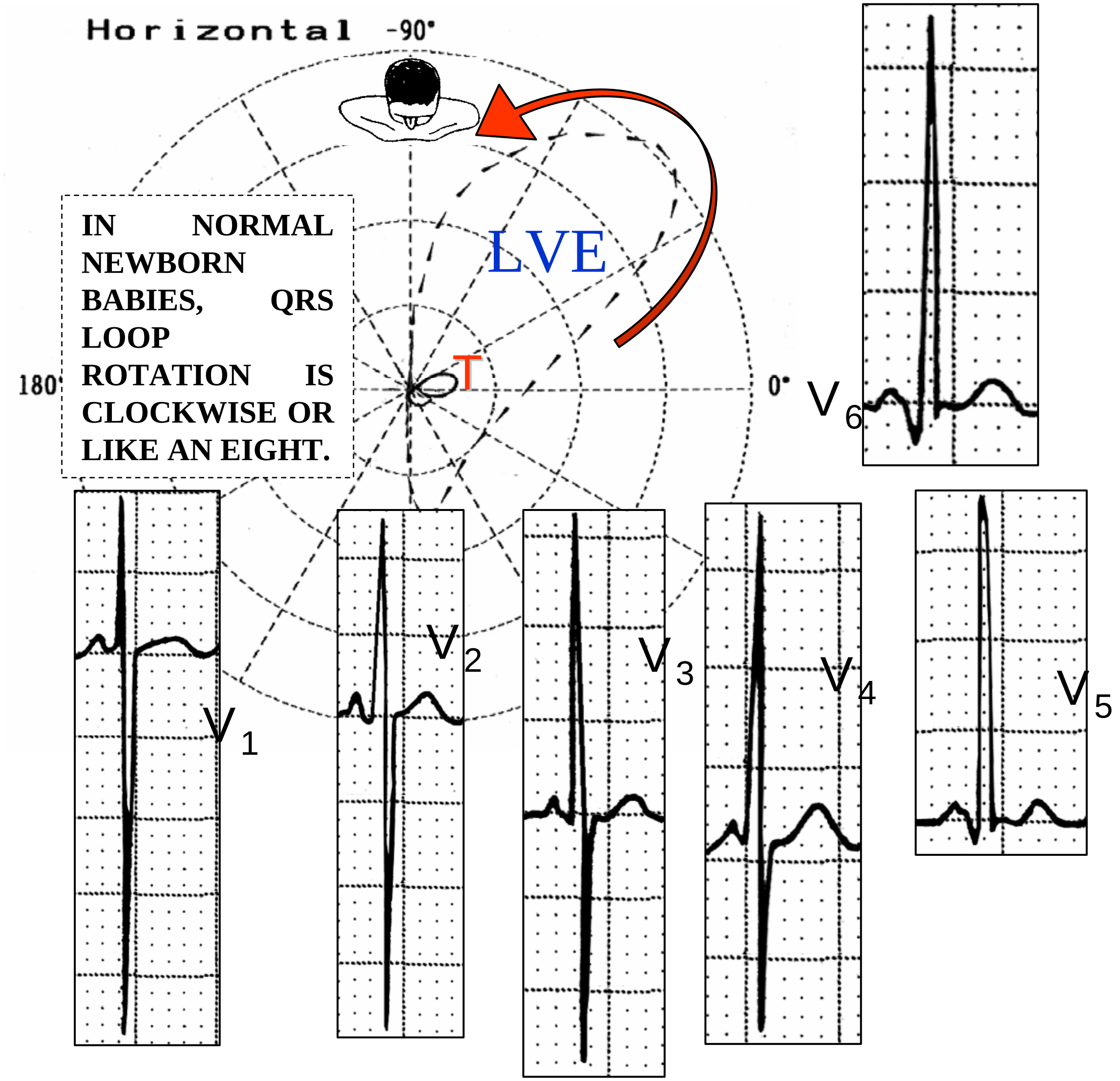
In normal newborn babies, QRS axis is around +125° in the right inferior quadrant. AQRS to the right.

In this case, QRS axis is in – 40°.

Association of LVH + LAFB + cyanosis, highly suggestive of tricuspid atresia.

ECG/VCG of TA in a newborn baby, cyanotic, 20 days of life. LAFB + LVE + adult progression in precordial leads. Typical LAFB in the FP and typical diastolic LVH/LVH in the HP.

ECG/VCG CORRELATION IN HORIZONTAL PLANE

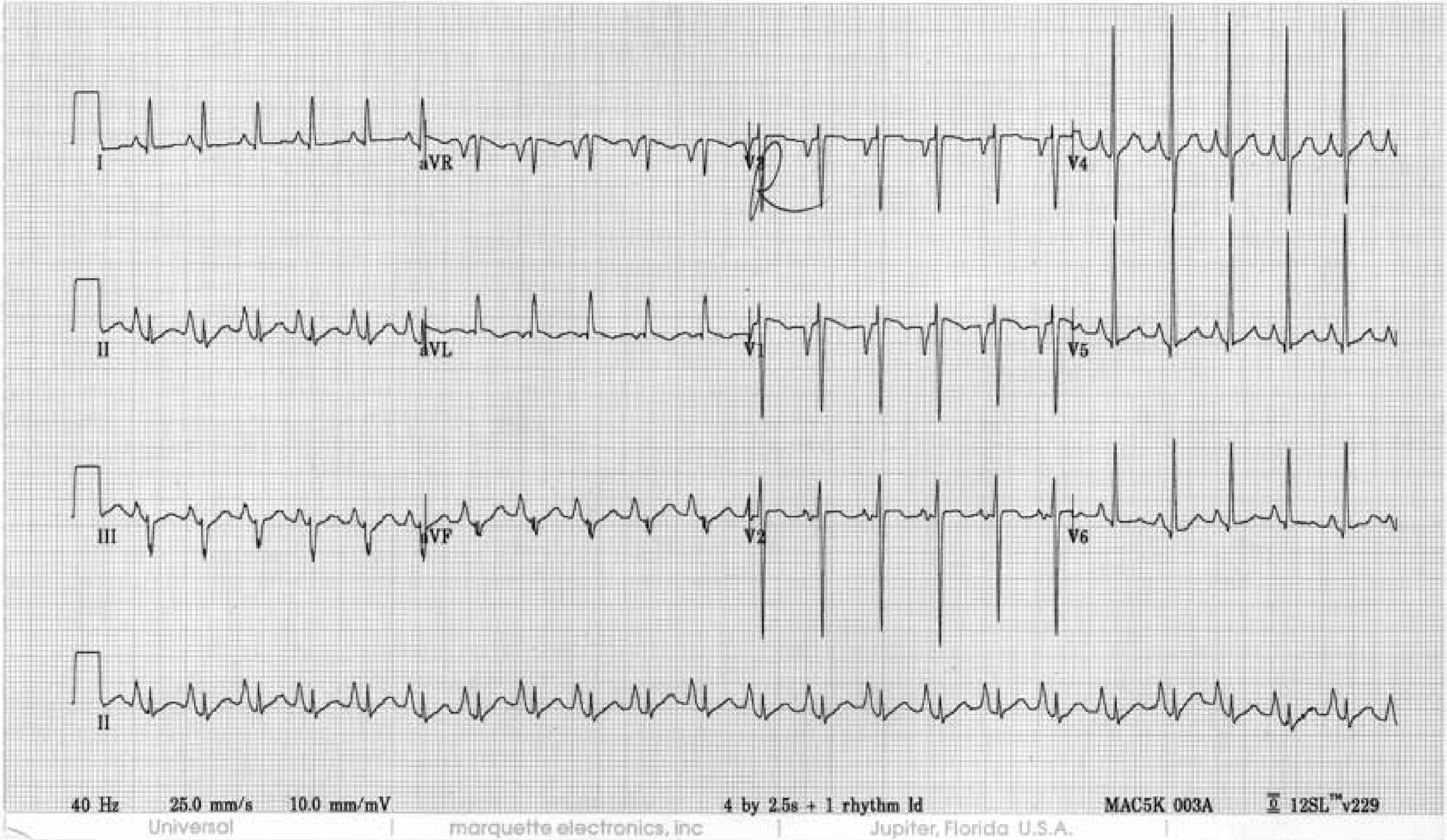


In the cases of TA without subpulmonary PS (Ic and IIc), the amplitude of R and S of V_1 is variable and V_6 presents deep Q followed by broad R.

In the HP of normal newborn babies, QRS loop is predominantly located in the right anterior quadrant: prominent R in V_1 and V_{4R} with R>S. We find here, left posterior shift of the QRS loop.
In our case, the QRS loop has an adult behavior: predominantly located in the left posterior quadrant.

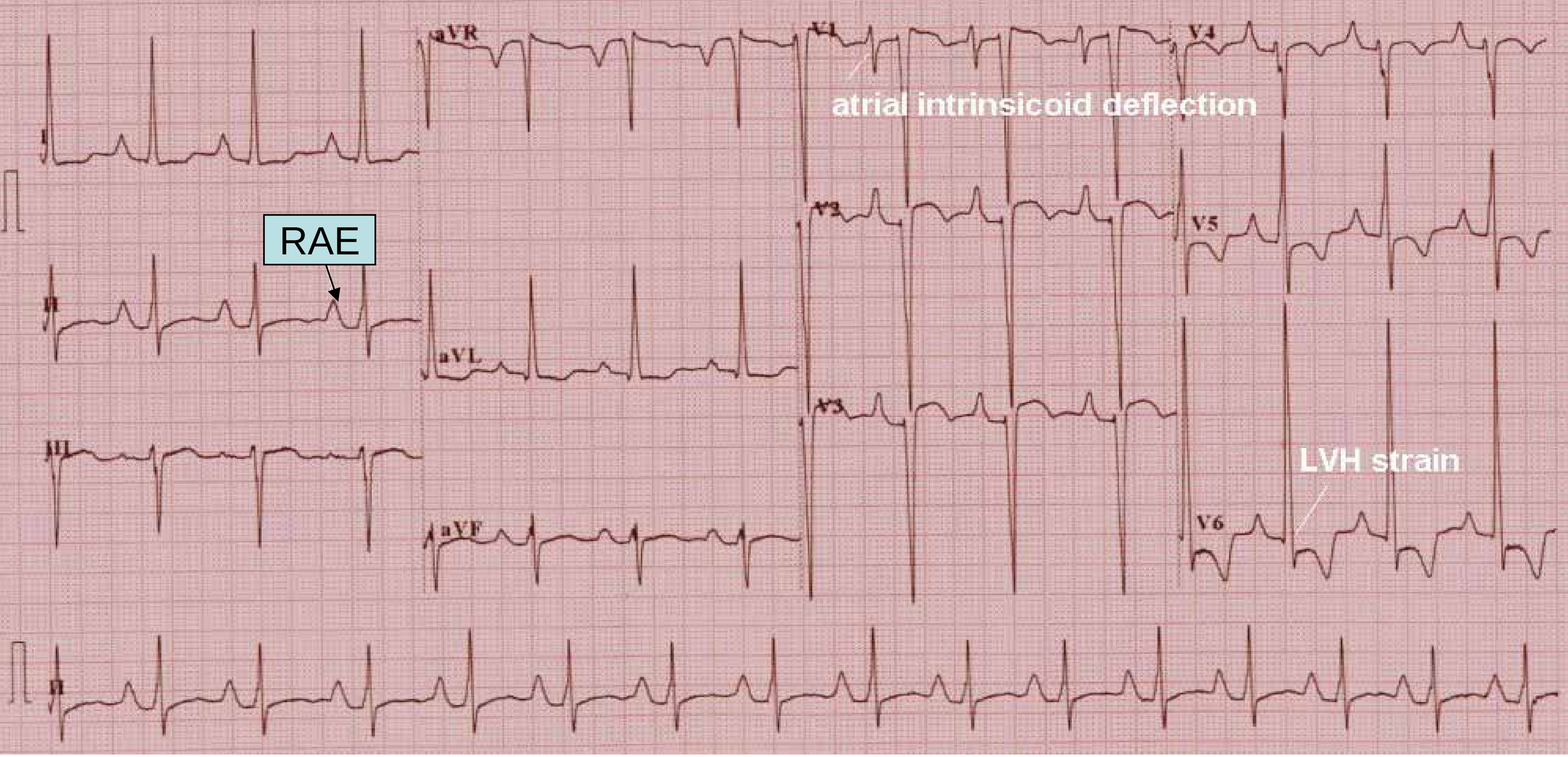
Adult progression of R/S in precordial leads is rare in newborn babies and when present, it suggests LVE/LVH and TA.

ECG/VCG of TA in a newborn baby, cyanotic, 20 days of life. LAFB + LVE/LVH + adult progression in precordial leads. Typical LAFB in the FP and typical diastolic LVE/LVH in the HP.



Sinus rhythm, with very tall and peaked P-waves (right atrial enlargement) P voltage >5 mm in lead II and profoundly negative in V1-V2. These types of P-waves are called giant P-waves or Himalayan P-waves and are indicative of a dilated right atrium due to a restrictive atrial communication.

QRS axis −15º, and an adult pattern of QRS progression over the precordial leads from V1 toV6. This pattern of QRS is characterized by absent right ventricular forces and well-developed left ventricular forces consistent with LVH/LVE.



Abnormally high voltage P waves in lead II. It is frequent in infants. Its height does not correlate well with atrial pressure or with the size of the atrial septal defect. There are descriptions of very high P waves
The biphasic P wave in V1 with sharp atrial intrinsicoid deflection (the sharp downward deflection from the peak of the P wave to the trough of the P wave) is a pseudo left atrial overload pattern, seen in right atrial overload. In true left atrial overload the atrial intrinscoid deflection is more slanting so that the negative component of the P wave is almost U shaped rather than the V shape in this case. The QRS axis is leftward with predominantly negative QRS in leads III and aVF.(-20º)
LVH with strain pattern is seen in lateral leads with tall R waves, ST segment depression and T wave inversion.

Medical management after palliation

The management issues in tricuspid atresia are similar to those in other cyanotic congenital heart defects and are discussed in TA.(1)

Hemoglobin should be periodically measured, and anemia and polycythemia, when present, should be treated.

Patients should receive antibiotic prophylaxis before undergoing any bacteremia-producing surgery or procedures.

The risks of stroke and brain abscess are similar to those in other cyanotic heart defects. When such a problem develops, appropriate neurologic or neurosurgical consultation and treatment is indicated.(1)

Routine well-child care, including immunizations, by the primary care physician is suggested. Administration of polyvalent pneumococcal vaccine and influenza vaccine and immunization against respiratory syncytial virus should be considered.(1)

Issues such as physical and emotional development, genetic counseling, vocational training and rehabilitation,[74] pregnancy, and contraception are addressed similarly to those in other cyanotic heart defects.[75]

The development of hyperuricemia, gout, and uric acid nephropathy in adolescents and adults with long-standing cyanosis and polycythemia is similar to that in other cyanotic heart defects.(1)Timely palliative and corrective surgery may prevent such complications.

1. *Rao PS, Covitz W, Chopra PS. Principles of palliative management of patients with tricuspid atresia. In: Rao PS, ed. Tricuspid Atresia. 2nd ed. Mount Kisco, NY: Futura; 1992:297-320.*
2. *Lichtman SW, Caravano M, Schneyman M, et al. Successful outpatient cardiac rehabilitation in an adult patient post-surgical repair for tricuspid valve atresia and hypoplastic right ventricle: a case study. J Cardiopulm Rehabil Prev. Jan-Feb 2008;28:48-51.*
3. *Strong WB, Morera JA, Rao PS. Sexuality, contraception and pregancy in patients with cyanotic congenital heart disease with special reference to tricuspid atresia. In: Rao PS, ed. Tricuspid Atresia. 2nd ed. Mount Kisco, NY: Futura; 1992:415-27.*

The prognosis for patients with AT and other complex congenital cardiac defects with one functioning ventricle has improved because of the advent of physiologically corrective surgery for TA and its modifications. However, such procedures are usually restricted to patients older than 1 year, though patients with TA are symptomatic in the neonatal period or early infancy. Palliation should be performed to allow infants to reach the age and weight requirements for correction. As a consequence, the objective of any management plan is not only to provide symptomatic relief but also to preserve, protect, and restore the anatomy (with good-sized and undistorted pulmonary arteries) and physiology (normal pulmonary artery pressure and preserved left ventricular function) to normal so that a corrective procedure can be safely performed when the patient reaches an optimal age and weight.(1)

Management at presentation

Medical management during the process of identification, transfer to a pediatric cardiology center, initial workup, and cardiac catheterization (if needed) and during and after palliative surgery or procedures includes maintenance of a neutral thermal environment, normal acid-base balance, normoglycemia, and normocalcemia with appropriate monitoring and correction, if necessary.(2;3) Unless associated pulmonary parenchymal pathology is present, the fraction of inspired oxygen (FIO2) administered should be no more than 0.4.

1. *Rao PS, Covitz W, Chopra PS. Principles of palliative management of patients with tricuspid atresia. In: Rao PS, ed. Tricuspid Atresia. 2nd ed. Mount Kisco, NY: Futura; 1992:297-320.*
2. *Rao PS. Management of neonate with suspected serious heart disease. King Faisal Spec Hosp Med J. 1984(4);209.*
3. *Rao PS. Principles of management of the neonate with congenital heart disease neonatology Today. 2007;2:1-10.*

Neonates who have low arterial PO2 and O2 saturation and ductal-dependent pulmonary blood flow should receive an intravenous infusion of PGE1 0.03-0.1 mcg/kg/min to open the ductus arteriosus or to maintain its patency.(1, 2;3] This is followed by an aortopulmonary shunt.

In the infant who presents with signs of congestive heart failure (type Ic or IIc), anticongestive therapy with digoxin, diuretics, and afterload reduction should be promptly given.(4) Considerations pertaining to pulmonary artery banding are reviewed in Surgical Care.

In patients with severe aortic coarctation, which is particularly observed in those with type II disease, ductal dilation with an infusion of PGE1 may improve systemic perfusion.(2) Surgical repair of the coarctation should follow. Some cardiologists use balloon angioplasty to relieve the aortic obstruction.

If interatrial obstruction is present, it should be relieved by means of balloon atrial septostomy. On occasion, blade or surgical septostomy is necessary.(5)

For patients presenting after infancy, the treatment approach is similar to that described above, except that PGE1 is not effective in opening the ductus.

1. *Freed MD, Heymann MA, Lewis AB, et al. Prostaglandin E1 infants with ductus arteriosus-dependent congenital heart disease. Circulation. Nov 1981;64:899-905.*
2. *Rao PS, Covitz W, Chopra PS. Principles of palliative management of patients with tricuspid atresia. In: Rao PS, ed. Tricuspid Atresia. 2nd ed. Mount Kisco, NY: Futura; 1992:297-320.*
3. *Rao PS. Principles of management of the neonate with congenital heart disease neonatology Today. 2007;2(8):1-10.*
4. *Rao PS. Principles of management of the neonate with congenital heart disease neonatology Today. 2007;2:1-10.*
5. *Rao PS. Transcatheter blade atrial septostomy. Cathet Cardiovasc Diagn. 1984;10:335-342.*

Catheter intervention

In the neonate, obstruction at the level of the atrial septum may be treated with conventional Rashkind balloon atrial septostomy.(1) In infants and children, the interatrial septum may be too thick to be torn with balloon septostomy; therefore, Park blade septostomy should precede the Rashkind procedure.(2)

In most patients, obstruction to pulmonary blood flow is at the VSD level or in the subpulmonary region. In some patients, the obstruction is at the pulmonary valve. In such patients, balloon pulmonary valvuloplasty may be useful in improving pulmonary blood flow and oxygen saturation.(3)

If progressive cyanosis develops after a previous Blalock-Taussig shunt and if the hypoxemia is due to a stenotic shunt, balloon dilatation may be used to improve oxygen saturation.(4;5) However, if the patient is of sufficient size and age to undergo a bidirectional Glenn procedure, this procedure should be performed instead of balloon angioplasty of a narrowed Blalock-Taussig shunt.

If severe aortic coarctation is present, particularly in patients with TA type II, balloon angioplasty may be useful in relieving aortic obstruction and may help achieve better control of congestive heart failure.(4)

If clinically significant branch pulmonary artery stenosis is present before bidirectional Glenn or Fontan conversion or after a Fontan procedure is performed, balloon angioplasty or placement of intravascular stents is recommended.(6)

1. *Rashkind W, Waldhausen J, Miller W, et al. Palliative treatment in tricuspid atresia. Combined balloon atrioseptostomy and surgical alteration of pulmonary blood flow. J Thorac Cardiovasc Surg. Jun 1969;57:812-818.*
2. *Rao PS. Transcatheter blade atrial septostomy. Cathet Cardiovasc Diagn. 1984;10:335-342.*
3. *McCredie RM, Swinburn MJ, Lee CL, Warner G. Balloon dilatation pulmonary valvuloplasty in pulmonary stenosis. Aust N Z J Med. Feb 1986;16:20-23.*
4. *Rao PS, Levy JM, Chopra PS. Balloon angioplasty of stenosed Blalock-Taussig anastomosis: role of balloon-on-a-wire in dilating occluded shunts. Am Heart J. Nov 1990;120:1173-1178.*
5. *Rao PS. An approach to the diagnosis of cyanotic neonate for the primary care provider. Neonatology Today 2007. 2007;2:1-7.*
6. *Rao PS. Stents in the management of congenital heart disease in pediatric and adult patients. Indian Heart J. Nov-Dec 2001;53:714-730.*

Development of aortopulmonary collateral vessels has been increasingly observed in recent studies. Before the final Fontan conversion, occlusion of these vessels in the catheterization laboratory, usually by means of coil embolization, is recommended to reduce left ventricular volume overloading and, probably, the duration of chest-tube drainage.

After a Fontan procedure, some patients may have recurrent pleural effusion, liver dysfunction, plastic bronchitis or protein-losing enteropathy. In these patients, rule out obstructive lesions in the Fontan circuit, then puncture of the atrial septum by using a Brockenbrough technique followed by static balloon atrial septal dilatation or stent implantation may be beneficial.

Patients who undergo a fenestrated Fontan operation or who have a residual atrial defect despite correction may have clinically significant right-to-left shunting that causes severe hypoxemia. These residual atrial defects may be closed by using transcatheter techniques.(1;2;3;4)

Some patients may develop systemic venous–to–pulmonary venous collateral vessels following Fontan operation, causing arterial desaturation. These vessels should be defined and closed by coils, plugs, or devices, depending on the size, location, and accessibility.(5)

1. *Rao PS, Chandar JS, Sideris EB. Role of inverted buttoned device in transcatheter occlusion of atrial septal defects or patent foramen ovale with right-to-left shunting associated with previously operated complex congenital cardiac anomalies. Am J Cardiol. Oct 1 1997;80:914-921.*
2. *Goff DA, Blume ED, Gauvreau K, et al. Clinical outcome of fenestrated Fontan patients after closure: the first 10 years. Circulation. Oct 24 2000;102(17):2094-2099.*
3. *Boudjemline Y, Bonnet D, Sidi D, et al. [Closure of extrocardiac Fontan fenestration by using the Amplatzer duct occluder]. Arch Mal Coeur Vaiss. May 2005;98:449-454.*
4. *Rothman A, Evans WN, Mayman GA. Percutaneous fenestration closure with problematic residual native atrial septum. Catheter Cardiovasc Interv. Oct 2005;66:286-90.*
5. *Sugiyama H, Yoo SJ, Williams W, et al. Characterization and treatment of systemic venous to pulmonary venous collaterals seen after the Fontan operation. Cardiol Young. Oct 2003;13:424-430.*

van den Bosch et al (1) studied the mortality, morbidity, and quality of life in our adult Fontan patients.
The authors examined 36 who underwent a Fontan procedure and were being seen in an adult outpatient clinic by using ECG, exercise testing, and echocardiography.
Quality of life was assessed by the Short Form 36 questionnaire.
The mean follow-up period was 15 years
28% of the initial 36 patients, 10 died after the Fontan operation and 1 patient underwent cardiac transplantation.
Reoperations were performed in 21 patients (58%), and the most common reason was revision of the Fontan connection.
Sustained supraventricular tachycardia was observed in 20 patients (56%) with an increased incidence of arrhythmias with longer follow-up.
Thromboembolic events were detected in 9 patients (25%), 5 of whom had adequate anticoagulant levels at the time of event. The thromboembolic event was fatal for 3 patients.
A total of 195 hospital admissions (mean 3.8 +/- 2.7, range 1 to 13) was recorded.

The authors found high mortality and very high morbidity in adult patients after the Fontan operation. In particular, reoperations, arrhythmias, and thromboembolic events compromised quality of life.

1. *van den Bosch AE, Roos-Hesselink JW, Van Domburg R, Bogers AJ, Simoons ML, Meijboom FJ. Long-term outcome and quality of life in adult patients after the Fontan operation. Am J Cardiol. 2004 May 1;93:1141-1145.*

Ablation of atrial tachycardia (AT) occurring late after cardiac surgery for congenital heart disease can be challenging due to the complexity of the arrhythmogenic substrate.
Tops et al (1) performed an ablation procedure in a Fontan patient with an AT using a new approach combining 3D electroanatomical mapping with multislice computed tomography (MSCT). This technique visualizes the position of the catheter in relation to the endocardium, thereby improving delineation of scar tissue. The AT had a focal origin located between areas of scar tissue and was successfully ablated at the earliest activated site. Ablation of complex arrhythmias can be facilitated by fusion of electroanatomical map and MSCT.

1. *Tops LF, de Groot NM, Bax JJ, Schalij MJ. Fusion of electroanatomical activation maps and multislice computed tomography to guide ablation of a focal atrial tachycardia in a fontan patient. J Cardiovasc Electrophysiol. 2006 Apr;17:431-434.*