

Corrected transposition of the great arteries

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Abstract

Corrected transposition of the great arteries is one of the most fascinating entities in congenital heart disease. The apparent corrected condition is only temporal. Over time, most patients develop systemic heart failure, even in the absence of associated lesions. With current imaging studies, precise visualization is achieved in each case though the treatment strategy remains unresolved. In asymptomatic patients or cases without associated lesions, focalized follow-up to assess systemic ventricular function and the degree of tricuspid valve regurgitation is important. In cases with normal ventricular function and mild tricuspid failure, it seems unreasonable to intervene surgically. In patients with significant associated lesions, surgery is indicated. In the long term, the traditional approach may not help tricuspid regurgitation and systemic ventricular failure. Anatomical correction is the proposed alternative to ease the right ventricle overload and to restore the systemic left ventricular function. However, this is a prolonged operation and not without risks and long-term complications. In this review the clinical, diagnostic, and therapeutic aspects are overviewed in the light of the most significant and recent literature. (Gac Med Mex. 2016;152:357-65)

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KEY WORDS: Corrected transposition of the great arteries. Double discordance. Anatomic correction. Double switch.

Introduction

Corrected transposition of the great arteries is one of the most fascinating entities in congenital heart disease. When a doctor sees for the first time a patient or a heart where the morphological right atrium is connected with the left ventricle (LV) and this in turn with the pulmonary artery, while the left atrium is communicated with the right ventricle (RV) and this in turn with the aorta, he/she cannot be less than surprised. First, due to the magnitude of alterations in the connections of cardiac segments: we are facing the discordances

that characterizes this condition: an atrioventricular discordance combined with ventriculoarterial discordance, and second, because caval venous return flow is correctly carried to the lungs, whereas pulmonary veins-originating oxygenated blood reaches the aorta and systemic circulation without any problem (Fig. 1). This situation of apparently “corrected” transposition goes on with no problem at least for the first decades of life, as long as there are no associated lesions. Things are, of course, more complex. Most cases have associated lesions, which will determine the clinical presentation, and having the RV supporting systemic circulation carries, over time, ventricular failure in most cases.

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Date of reception: 04-03-2015

Date of acceptance: 06-04-2015

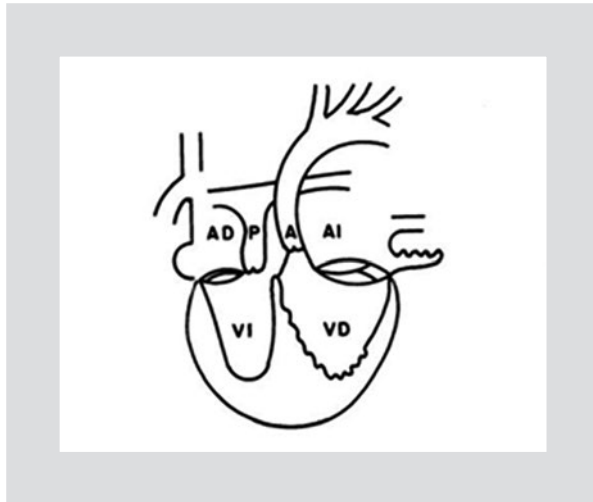


Figure 1. Drawing where the double discordance in situs solitus is visualized. The atria are recognized by the morphology of their auricles: the right one, broad-based, and the left one, narrow and with an irregular, scallop shape. The left ventricle with smooth endocardium and the right with thick trabecula. Observe that the great arteries do not cross but emerge in parallel, where the aorta appears left and straight. By virtue of the double discordance, circulations have a normal course. RA: right atrium; LV: left ventricle; LA: left atrium; P: pulmonary artery; A: aorta.

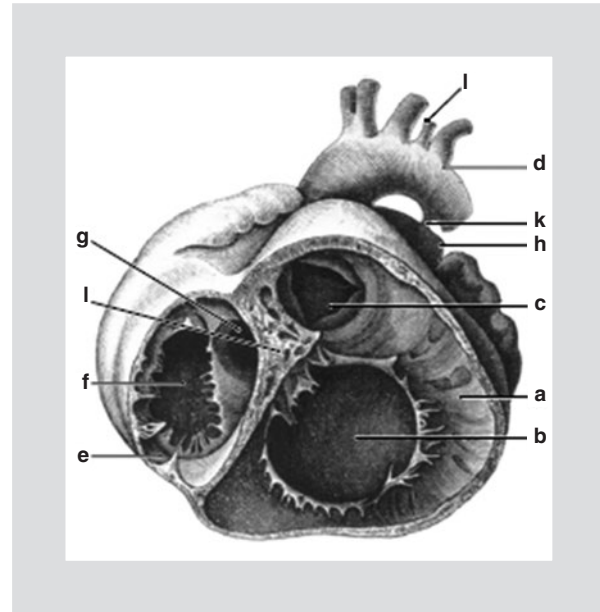


Figure 2. Original drawing of a corrected transposition in situs solitus made by Rokitansky in 1875. It is a cross-sectional cut at the level of the atrioventricular valves. The right ventricle (A) is observed to be dilated, (B) Tricuspid valve, supporting the aorta (C), (D) Aorta, (E) Left ventricle and left with normal size (F), and in relation to the pulmonary artery (G). The view is very much alike to that obtained with bi-dimensional echocardiography.

In this review, we will analyze all these aspects with more detail, with particular emphasis on clinical manifestations, diagnosis, prognosis and treatment.

History

The first known description of corrected transposition of the great arteries (CTGA) was made by baron von Rokitansky in 1875 with a very beautiful illustration (Fig. 2), where both discordances can be appreciated in situs solitus. Since then, an elevated number of CTGA cases and all type of associated lesions have been described and documented.

Nomenclature

In this article, when it reads LV, we always refer to the morphological left ventricle, the same way than when it reads RV, reference is being made to the morphological right ventricle, regardless of their spatial situation within the thorax.

On the other hand, although CTGA is referred to rather frequently as “L” transposition due to its location left to the aorta in the frontal projection, this is inaccurate, since situs inversus CTGA itself has the aorta at the right and other complex pathologies can have the

aorta in “L position”, such as the RV double outflow way. Therefore, the accurate form is to describe it as atrioventricular (AV) and ventriculoarterial discordance in situs solitus or inversus and then the associated lesions, without forgetting the position of the heart within the thorax, as we will address later.

Etiology

The etiology of this malformation is unknown; however, the L-loop during embryogenesis is known to be determinant in ventricular inversion and it is responsible for the presence of double discordance. There is predominance in the male gender in a 1.6 to 1 ratio.

Frequency

CTGA affects from 0.4 to 1% of all children with congenital heart disease¹⁻⁴ but it is likely to be underestimated, since those with no associated lesions usually have clinical manifestations at older ages and in some cases they may not show manifestations of this heart condition and die of other cause. In approximately 80 to 95% of the cases it occurs in situs solitus and in 5 to 20% in situs inversus⁵⁻⁷.

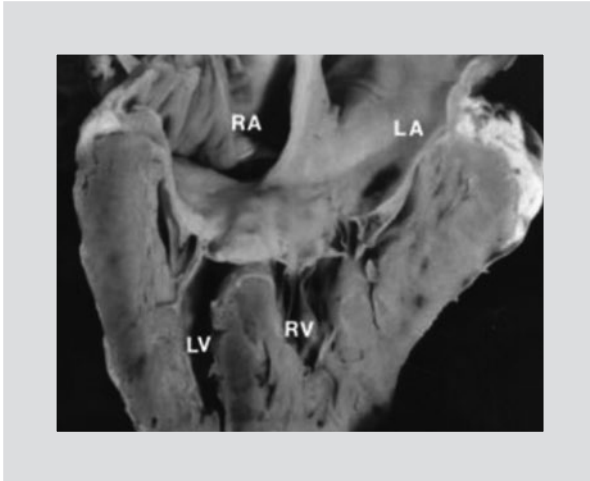


Figure 3. Specimen showing atrioventricular discordance and, as associated lesion, a complete atrioventricular septal defect can be observed. A common atrioventricular valve riding on the interventricular septum is appreciated. LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.

Anatomical characteristics and associated lesions

Notably, in most cases, the interventricular septum has a sagittal orientation with the ventricles at both sides in hearts that tend to mesocardia within the thorax (Fig. 3), but the cardiac shape can appear in levo- or dextrocardia^{8,9}.

The pulmonary ring is wedged onto the center of the heart between the mitral and tricuspid annulus and therefore the membranous septum has a larger area,

whereas the His bundle is longer, with a trajectory anterior to the infundibular septum that predisposes to atrioventricular block, as we will see in the clinical section^{10,11}. The aortic annulus, supported by the morphological right ventricle infundibulum is located up and to the right, giving rise to a straight, ascending and left aorta.

Practically any known associated lesion in patients with concordant hearts can occur in CTGA (Fig. 3); however, there are some lesions and combinations thereof that are characteristic: the combination of ventricular septal defect with pulmonary stenosis (Fig. 4) and tricuspid lesion with regurgitation is a characteristic triad of CTGA¹²⁻¹⁴.

If we analyze the lesions separately, the most common lesion is interventricular communication (IVC), which is present in at least 80% of cases¹⁵; usually, it is perimembranous, but it can be found at any site or it can be multiple. Pulmonary obstruction is observed in 40 to 70%¹⁶; characteristically, it is a subpulmonary obstruction by accessory tissue (Fig. 4), but it may be combined with stenosis at the valvular level. Pulmonary atresia has been found in 8% of cases¹⁷. Tricuspid anomalies are universal in autopsy cases and, clinically, at least one third of patients have significant tricuspid insufficiency¹⁸. Aortic insufficiency has been reported in approximately 30% of cases¹⁹. Of course, CTGA is also accompanied by interatrial communication (IAC) or patent ductus arteriosus (PDA), either in isolated form or associated with important lesions such as IVC, pulmonary stenosis and tricuspid insufficiency.

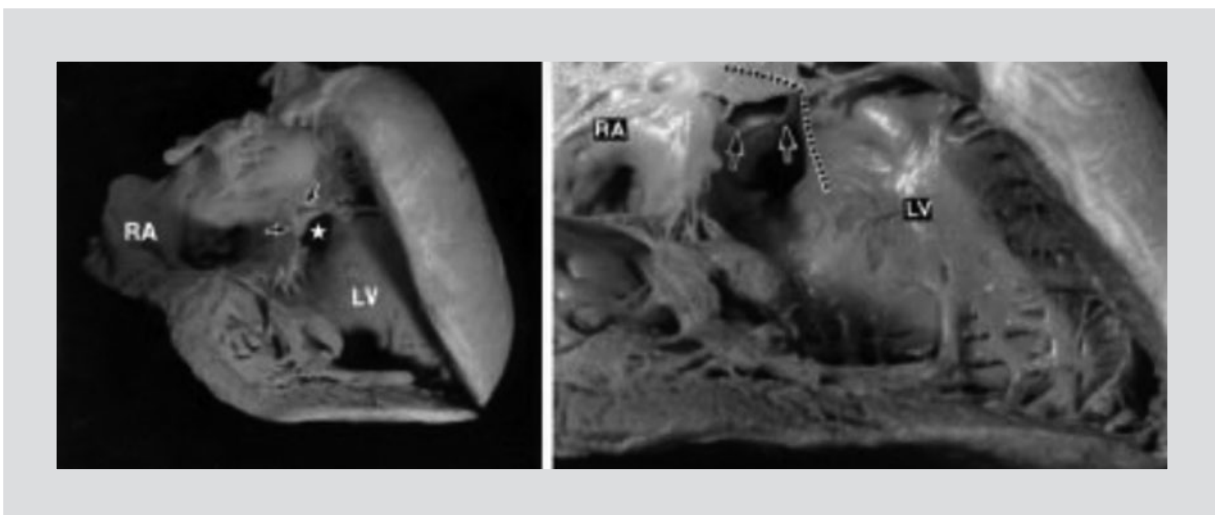


Figure 4. A: specimen with CTGA. This view corresponds to the morphological left ventricle. Smooth septum and fine trabeculae are appreciated. Observe a restrictive subpulmonary interventricular communication (asterisk). **B:** closer view of the same specimen. Subpulmonary obstruction pointed by vertical arrows and, in dotted line, the route of the conduction tissue across the anterior and superior margin of the septal defect. RA: right atrium; LV: left ventricle (adapted from Alva C. et al.)²⁰

Coronary circulation originates in the sinuses of Val-salva posterior to the aorta. In situs solitus, the left coronary artery arises from the right one and irrigates the LV with its usual pattern, whereas the right coronary artery arises from the left one and is normally distributed across the RV. Different coronary anomalies have been described, such as single coronary artery²⁰.

Pathophysiology

The pathophysiology of associated lesions is the same than in cases where there is no double discordance. For example, patients with significant pulmonary stenosis and large IVC and double discordance behave the same as in a tetralogy of Fallot without CTGA. However, higher frequency of AV block (longer bundle of His) that progresses over time is exclusive to CTGA when compared with populations without CTGA. Similarly, systemic ventricular failure over time is characteristic of this entity, as we will immediately see in the natural history section.

Natural history

In fetal life

Prenatal diagnosis is feasible by means of echocardiogram (ECHO) in experienced hands. Associated lesions are the same than after birth. Some fetuses die due to the severity of lesions and interruption of pregnancy is decided in other cases^{21,22}.

CTGA without associated lesions

This group represents one of the most fascinating and interesting groups studied by congenital heart disease investigators. Between 1 and 16% of patients with double discordance are estimated not to have associated lesions^{15,23}; however, there could be more, since many of them never receive attention for heart conditions because of being asymptomatic and others die from other causes without having been studied. We know that survival without associated lesions is much better than with their presence; there are reports of patients with CTGA without associated lesions alive at the eightieth and ninetieth decades of life. However, most experience RV systemic ventricular failure from the fourth decade onwards and 25% have heart failure at 45 years of age, as compared with 67% of patients with CTGA with associated lesions¹⁹.

A large number of patients without associated lesions initiate their deterioration with the presence of tricuspid insufficiency, which determines volume overload, annular dilatation and further insufficiency. This vicious circle precipitates ventricular failure of the systemic ventricle. The background explanation of this is an anatomical factor: the RV ventricle architecture is not adequately prepared to bear the burden of systemic circulation all life-long. What we cannot explain is why some patients can reach older ages without ventricular failure.

The first manifestations of patients with CTGA without associated lesions are usually signs and symptoms of low output due to advanced AV block. The need for pacemaker is a common event. Presbitero observed 18 patients without associated lesions, out of whom 7 (38.8%) required pacemaker in an average follow-up of 10 years²⁴.

Clinical presentation of CTGA with associated lesions

From a general perspective, clinical presentation is that of an associated lesion without double discordance; for example: a large IVC will occur with manifestations of heart failure 4 weeks after birth when the drop of pulmonary resistances causes for the left-to-right shortcut to be significant. The combination of IVC and pulmonary stenosis will be recognized as a tetralogy of Fallot, i.e., significant cyanosis with pulmonary oligohemia. When tricuspid regurgitation is added to the IVC and pulmonary stenosis dyad (a common triad in CTGA), cyanosis has signs and symptoms of "mitral" insufficiency added, which may cause manifestations of pulmonary congestion. There are, of course, cases with isolated PDA or IAC, where clinical manifestations are those produced by each one of these entities.

Pregnancy can precipitate heart failure manifestations due to volume overload, in a patient with tricuspid regurgitation, expressed as mitral insufficiency. Dyspnea with heavy exertion can also be the first manifestation in young adults when systemic ventricular failure becomes apparent. In all these cases, clues to suspect CTGA must be considered.

Clues to suspect that a particular lesion has an underlying CTGA

Clinically, just with stethoscope it is difficult; however, auscultation of heart sounds located at the center of the chest or clearly at its right side, associated with

an accentuated 2nd “pulmonary” sound, which actually is the closure of the aortic valve anterior and left-located, should be considered as elements for suspicion. This suspicion is reinforced when the chest X-ray is analyzed and a cardiac silhouette in the meso- or dextrocardia position is observed²⁻²⁴, with morphology tending to be square-shaped and with the straight and high left profile that corresponds to the aorta.

Clues in electrocardiogram

Most CTGA cases are found in atrial situs solitus and in them, ventricular septal depolarization is inverted, i.e., from right to left, with this determining for Q-waves to be observed in the right precordials and to disappear in the left ones (opposite to normal); Qs patterns can also be observed in DII and AVF leads with electrical axis to the left. This is not observed in patients with situs inversus. AV conduction defects are common and progressive; only 40% of 7-year old children with CTGA have normal conduction²⁵, and in adulthood, 60% of cases have some conduction defect^{26,27}. Holter monitoring may be indicated to assess for block and possible arrhythmias. Of course, associated lesion(s) can produce different types of overload associated with the described data. These clues, together to clinical and radiological findings, give support to strong suspicion of double discordance that should be corroborated with ECHO.

ECHO

Applying the segmental analysis methodology in the study of these patients is imperative. First, it is essential to establish the atrial situs, which can be inferred from a cross-sectional slice at the abdominal level, by observing the relationship of the vena cava and the aorta with regard to the vertebral column. Atrioventricular connections are established based on anatomical criteria: in situs solitus, the morphologic LV will be recognized by a smooth endocardium, only a pair of papillary muscles and one bicuspid valve, the mitral valve (Fig. 5). In the absence of IVC, mitral valve implantation level is higher than that of the tricuspid. This ventricle is observed to be connected to the right atrium and located at the contralateral's left, whereas the left atrium will be observed to be connected to the anatomical right ventricle located at the right, which is recognized by its thick trabecula, presence of the moderator band, implantation of a papillary muscle to the septum and the tricuspid valve (Fig. 5)²⁸⁻³⁰. The aortic valve, together with the aorta, is appreciated almost

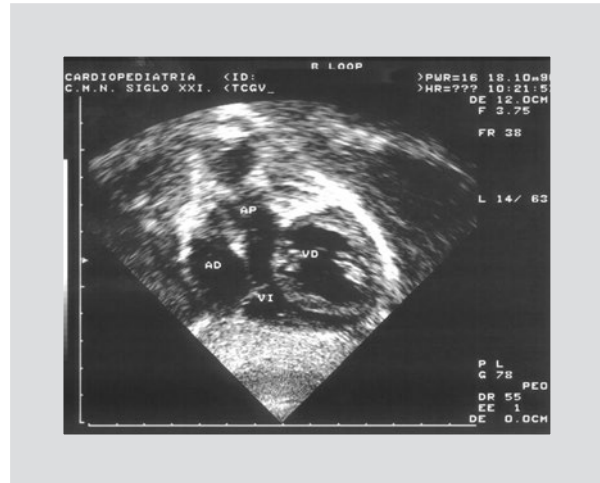


Figure 5. *Situs solitus CTGA in an infant. Posterior subcostal approach. The atrioventricular connection is appreciated where the right atrium (AD) connects to a thin wall left ventricle (VI), with the interventricular septum protruding to its interior. The LV is connected with the pulmonary artery (AP) without stenosis. The right ventricle is hypertrophic and rounded.*

always being held by an anterior infundibulum. In the absence of IVC, tricuspid implantation in the septum is lower than mitral implantation³⁰.

Associated lesions by ECHO

As previously mentioned, pulmonary valve-related perimembranous IVC is quite common, same as the recognition of pulmonary obstruction, which generally is sub-pulmonary and caused by accessory tissue. Continuous Doppler specifies the obstruction gradient, as well as the gradient between ventricles. It is also quite common observing tricuspid valve alterations, ranging from moderate dysplasia to clear displacement towards the ventricle interior, as in Ebstein anomaly. In all cases, the magnitude of regurgitation should be quantified, and ventricular function measured in both ventricles. Morphological left ventricle function will hardly be abnormal in non-operated cases; in contrast, different degrees of dysfunction are likely to be found in the systemic ventricle, often directly associated with the degree of tricuspid insufficiency³⁰. In some adults with an inadequate window, transesophageal ECHO may be necessary.

Cardiac catheterization and angiocardiography

Currently, in most cases, ECHO is sufficient to establish the diagnosis and program surgery; however, there are precise indications for cardiac catheterization

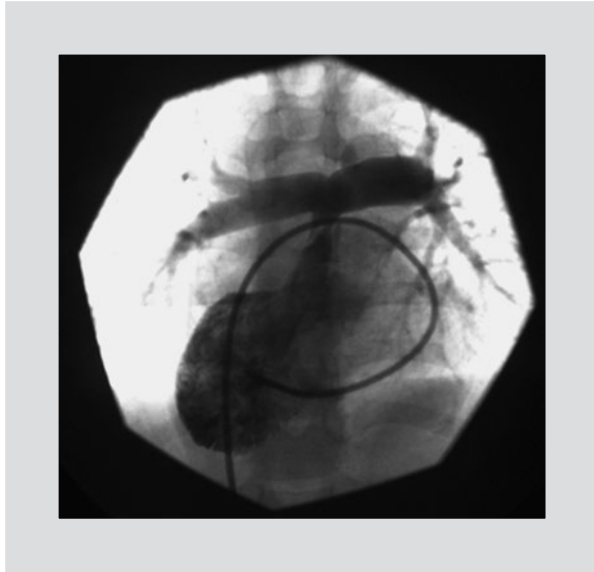


Figure 6. Angiocardiogram of a patient with CTGA in frontal projection. The catheter ascends to the right through the inferior vena cava, turns to the left in the atria, through a foramen ovale, passes through the tricuspid valve to the right ventricle and, through interventricular communication, reaches the left ventricle (LV), where the injection is applied. The LV is recognized by its fine trabecula and play card shape. A pulmonary valvular stenosis with a short stem and good branches of the pulmonary artery are observed. The L-shaped aorta is faintly appreciated.

and selective angiography (Fig. 6): In cases with pulmonary atresia associated with aortopulmonary collateral arteries, selective angiographies of collaterals is the best way to visualize them when in a patient, beyond childhood, there is severe pulmonary arterial hypertension and measuring the pulmonary resistance pressure units is necessary, or in patients with a previous history of fistula, and in cases where LV training with banding has been performed and anatomical correction is planned. Visualization of the coronaries is also important when double switch operation is planned.

Computed tomography and magnetic resonance imaging

Both these non-invasive studies are most useful, when they are available, to assess the ventricular function in particular and postoperative conditions of patients who evolve poorly. Anesthesia may be required in small children, with its non-invasive nature thereby disappearing.

Differential diagnosis

Some entities must be disregarded, such as ambiguous AV connections where there is atrial isomerism and the anatomical left ventricle is located at the right,

or rare cases with isolated ventricular inversion, where there is AV discordance, but ventriculoarterial connections are concordant. In all of these situations, systematic and careful application of echocardiographic segmental analysis should establish the right diagnosis.

Indications for surgery

In the field of complex congenital heart conditions, sometimes it is difficult to decide when and how to intervene; CTGA is one of them. Grouping the available approaches is convenient:

- Palliative procedures: In newborns and small infants with pulmonary stenosis or atresia with severe hypoxemia and hypoxic crisis, a systemic-pulmonary fistula solves the hypoxia. In infants with heart failure and large septal defects, especially when associated with tricuspid regurgitation, pulmonary artery banding can control the heart failure and relieve the tricuspid insufficiency by reducing pulmonary flow. Pacemaker is indicated in patients with symptomatic complete AV block.
- Complete repairs
 - Physiological, classical, traditional or conventional correction

Until 25 years ago, and for several decades, surgical approach for these patients was that of associated lesions, i.e., septal defects closure, pulmonary stenosis relief or LV and pulmonary artery valved tube (Fig. 7) or tricuspid valve plasty. Surgical mortality went on gradually decreasing until a range of less than 5 to 20% was reached, depending on the associated lesions³¹⁻³⁶. Although initial evolution was satisfactory, in the mid- and long-term follow-up it became evident that, in spite of correctly repairing associated lesions, over time, most cases develop tricuspid regurgitation and right ventricular dysfunction, which become apparent as heart failure³³⁻³⁶. A multicenter study documented that, at 45 years of age, 67% of patients treated with the classical approach had insufficiency¹⁹. In summary, the traditional surgical approach demonstrated that, over time, tricuspid insufficiency and right ventricular failure were common complications associated with poor prognosis³⁷. This prompted the search for alternatives:

 - Anatomical correction, double switch

Ilbawi et al.³⁸, to solve the problem of systemic ventricular failure, initiated anatomical correction

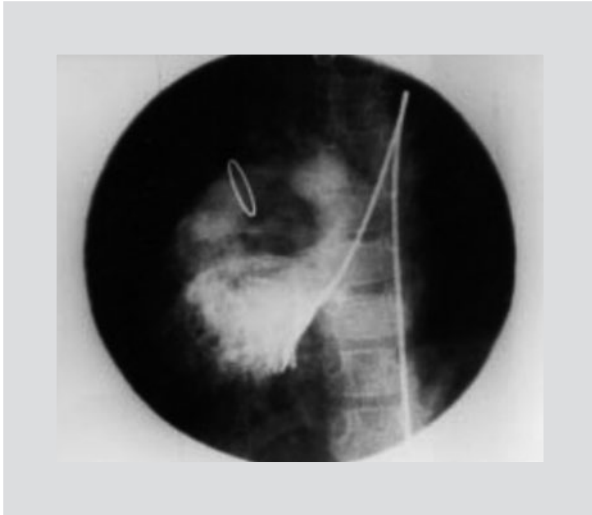


Figure 7. *Traditional or classical approach. Six-year old boy with CTGA, situs solitus, dextrocardia with severe pulmonary stenosis. Associated lesions were surgically treated: Ventricular Septal Defect (VSD) closure was made with a patch and a valved tube was placed communicating the left ventricle with the pulmonary artery (Rastelli). The control angiographic study shows, in the frontal projection, the catheter that was introduced via the femoral artery ascending through the aorta, travelling across the aortic arch (left and straight), entering the right ventricle and, through a small residual IVC, entering the LV, out of which the valved tube towards the pulmonary artery arises (adapted from Alva C. et al.)⁵.*

with the purpose to connect the LV with the aorta, and the RV with pulmonary circulation. Since then, two main approaches have been developed^{14,38-46}: in the presence of pulmonary stenosis or atresia, a switch is practiced at the atrial level: Senning or Mustard, and the aorta is reoriented with the LV through the IVC by means of a patch; in addition, a Rastelli tube is implanted to connect the RV with the pulmonary artery. In the cases with normal pulmonary valve and absence of obstruction of this outflow way, atrial switch is performed together with arterial switch to correct the ventricles' functions with their appropriate circulation and, finally, associated lesions are corrected. An important proportion of patients without both pulmonary stenosis and IVC cannot undergo double switch because the LV handles pulmonary artery low pressures and is not fitted to endure systemic pressure and, for this reason, it has to be trained by means of a previous intervention: pulmonary artery banding⁴⁵. The results of anatomical correction with training by means of banding have not been less favorable. The most important variable is the age at which the banding is practiced; in children older than 15 years it doesn't

seem recommendable planning an anatomical correction by means of pulmonary artery banding⁴⁶⁻⁴⁷.

Although anatomical correction fundamental goal is achieved, i.e., placing the LV under systemic circulation, these operations are complex and long-lasting and not free of surgical risk; therefore, they should be performed in centers with experience in both arterial and atrial switch. As one can imagine, in postoperative follow-up, problems related to the procedures have emerged: Need for valved tube replacement in cases with Rastelli, whereas in cases with arterial switch, supra-valvular stenoses and aortic insufficiency have been observed⁴⁸. In addition, obstructions at the level of atrial redirection and emergence of arrhythmias have been common complications to both procedures³⁹. Finally, postoperative AV block occurs both in the traditional approach and anatomical correction^{48,49}.

Specific situations

Bearing in mind the principle that each case should be individualized, we expose below some more specific recommendations.

In children or adults with CTGA with no associated lesions, or when these are so mild that they have no significant hemodynamical repercussion, such as a small IVC or a mild pulmonary stenosis, we consider management should be medical with periodic surveillance every 6 months with clinical, electrical and echocardiographic evaluation. We don't believe it is advisable for an asymptomatic patient to undergo pulmonary banding for LV training. In these cases, attention should be focused on tricuspid valve, systemic ventricular function and conduction tissue monitoring. This conservative management is based on the fact that asymptomatic individuals, especially children, have a good prognosis at least until the fourth decade of life. In adults, it is more complicated, since the prognosis is not so good but, given that we don't know which patients will remain asymptomatic for several years and which won't, and that anatomical correction bears risk, a preventive operation doesn't seem to be warranted. Of course, if any lesion is important in adulthood, intervention should be evaluated.

When an infant is symptomatic due to associated lesions, surgical intervention is indicated, as in cases without double discordance. For example: in a patient with IVC and severe pulmonary stenosis or pulmonary atresia, with hypoxia crisis, a first option is to practice a systemic-pulmonary fistula.

Traditional correction with IVC closure and pulmonary stenosis correction, either by relieving it or by implanting a valved tube, can also be chosen. The decision to perform the anatomical correction at the same time will depend on the experience of the center and the state of the tricuspid valve and the functioning of the morphological right ventricle. If there is ventricular dysfunction with tricuspid regurgitation, proceeding with anatomical correction is recommended. The reasoning is the following: with the traditional approach, the RV would be left functioning as the systemic ventricle and, since when pulmonary obstruction is relieved pulmonary flow increases; this will favor an increase in tricuspid regurgitation and right ventricle failure, with both leading to heart failure manifestations. This means performing atrial redirection together with LV-aorta connection by means of a hammock-like patch through the IVC and implanting a valved tube from the RV to the pulmonary artery, thus restituting LV systemic function and relieving the load from the RV by connecting it to the pulmonary circulation.

In children with broad IVC, without pulmonary stenosis, tricuspid insufficiency and right ventricular dysfunction, where both ventricles handle high pressures, double switch is indicated: at the atrial and arterial levels. The decision is complicated when the LV has low pressure because of lacking IVC or pulmonary stenosis. These cases require LV training with an intermediate operation: pulmonary artery banding; however, results have not been that good, particularly after 15 years of age. It is important to note that some cases with CTGS have marked hypoplasia of any of both ventricles; for this particular group of patients with ventricular hypoplasia, other options such as the hemi-Mustard and Fontan^{50,51} operation modifications have been proposed with encouraging results in the short and long-term.

In view of all the above, CTGA clearly remains a challenge, specially a therapeutic one. Comparative, prospective studies are required, as well as new techniques that reduce complications and offer better survival.

Conclusions

- CTGA is one of the most interesting congenital heart conditions and represents a separate population where, owing to double discordance, circulations are “corrected”.
- Most patients will experience tricuspid insufficiency and systemic ventricular failure in adulthood.

- Clinical presentation in minors is determined by associated lesions.
- There are clinical, electrical and radiological clues to suspect the diagnosis, but the most important is bearing it in mind.
- To this moment, there are not enough reasons to intervene asymptomatic individuals.
- Traditional surgical approach has the following advantages: vast experience, reduced surgical times and low mortality. Disadvantages: tricuspid insufficiency and right ventricular failure cannot be avoided.
- In turn, anatomical correction or double switch restitutes RV to pulmonary circulation and LV to systemic circulation, thus preventing tricuspid regurgitation and heart failure; however, it is a complex and prolonged procedure. Its most common complications include arrhythmias and venous obstructions. Aortic insufficiency and coronary obstructions are less common, but have been documented.
- Both approaches share AV conduction disorders and the need of valved tube replacement when there is pulmonary stenosis.
- Comparative long-term studies are needed as well as the promotion of new techniques.
- CTGA is an extraordinary entity that is far from being resolved. Patients cannot be considered as being cured and follow-up should be permanent.

Acknowledgements

The author thanks Héctor Alva Sánchez, PhD, for his support in the English language.

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