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Review Article

Overview of Transposition of Great Arteries in Newborn

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Abstract

Introduction: Complete transposition of the great arteries (D-TGA) is the most common cyanotic congenital heart defect in newborns, at least in Western countries. In this condition, the aorta arises from the right ventricle (RV) and the pulmonary artery (PA) arises from the left ventricle (LV). As the result, the normal anteroposterior relationship of the great arteries is reversed, so that the aorta is anterior to the PA (transposition), but the aorta remains to the right of the PA; thus, the prefix D is used for dextroposition. In levo-transposition of the great arteries (L-TGA, or congenitally corrected TGA), the aorta is anterior to and to the left of the PA; therefore, the prefix L is used. The atria and ventricles are in normal relationship. The coronary arteries arise from the aorta, as in a normal heart. Desaturated blood returning from the body to the right atrium (RA) flows out of the aorta without being oxygenated in the lungs and then returns to the RA. Therefore, tissues, including vital organs such as the brain and heart, are perfused by blood with a low oxygen saturation. Conversely, well-oxygenated blood returning to the left atrium (LA) flows out of the PA and returns to the LA. This results in a complete separation of the two circuits. The two circuits are said to be in parallel rather than in series, as in normal circulation. This defect is incompatible with life unless communication between the two circuits occurs to provide the necessary oxygen to the body. This communication can occur at the atrial, ventricular, or ductal level or at any combination of these levels. In the most frequently encountered form of D-TGA, only a small communication exists between the atria, usually a patent foramen ovale (PFO). The newborn is notably cyanotic from birth and has an arterial oxygen saturation of 30% to 50%. The low arterial PO2, which ranges from 20 to 30 mm Hg, causes an anaerobic glycolysis, with resulting metabolic acidosis. Hypoxia and acidosis are detrimental to myocardial function. The normal postnatal decrease in pulmonary vascular resistance (PVR) results in increased pulmonary blood flow (PBF) and volume overload to the LA and LV. Severe hypoxia and acidosis (with a resulting decrease in myocardial function) and

volume overload to the left side of the heart cause CHF during the first week of life arterial switch operation is usually done to correct the arterial transpositions.

The Aim of Work: The review outlines the pathophysiology, clinical manifestations, etiology, diagnostic methods and management of transposition of great arteries.

Methodology: The review is comprehensive research of PUBMED from the year 1986 to 2018.

Conclusion: There has been a massive improvement seen in the survival of children with the transposition of arteries. Successful surgery is seen in over 95% of patients if no unusual risk factors are involved. There is a normal reconstruction of the heart, both anatomically and functionally, to restore the heart to a normal condition. The arterial switch operation usually results in a normal ventricular function and no heart rhythm abnormalities. ASO is the surgery of choice to be performed in infants who are diagnosed early. Satisfactory therapeutic effects can be achieved by strengthening intraoperative procedures and post-operative care.

Keywords: *Congenital; Heart Defect; Porto-systemic Shunt; Arteries*

Introduction

The transposition of great arteries (TGA) is often called complete transposition is a cardiac malformation seen congenitally, which has features of atrioventricular concordance and ventriculoarterial discordance [1]. This condition accounts for about 5 - 7% of the congenital heart diseases which corresponds to 20 - 30 per 100 000 live births. The male to female ratio is 1.5 to 3.2:1 [2]. Although it is a life-threatening malformation in neonates, it can be repaired completely. Early detection diagnosis and management prenatally can reduce the morbidity and mortality of the patient. This was first described in 1797 by Mathew Baillie, but only in 1814 did the term transposition get applied, which means the placement (position) of the aorta and pulmonary trunk across (trans) the ventricular septum [3].

In this congenital malformation, the aorta arises from the morphological right atrium and ventricle, and the pulmonary trunk emerges from the morphological left atrium, which is connected to the left ventricle [3]. Physiologically uncorrected transposition is described by the term complete transposition. Commonly the prefixes a-, d- and l- transposition is used which gives a description of the spatial relationship between the aorta and pulmonary trunk. Like in d-transposition, the aortic valve lies on the right side of the pulmonary valve. l-transposition refers to a condition when the aorta usually lies on the left in congenitally corrected hearts. a-transposition refers to when the pulmonary trunk is related to the anterior position of the aortic valve. Other non-cardiac malformations may be associated with this cardiac lesion [1].

The anatomical correction by arterial switch operation (ASO) brings about a favorable outcome; this has become the preferred option for treating TGA. It is commonly suggested for infants to undergo the ASO surgery within two weeks of their birth. However, the specific timing of the disease still remains a controversy [4].

Pathology, pathophysiology, and aetiology

Retention of the usual configuration of the atriums and ventricles are seen in hearts with transposition of arteries. The tricuspid valve is separated from the aortic valve by a right-sided subaortic infundibulum, and there is a presence of fibrous continuity between the mitral and pulmonary valve. Even though Coronary arteries' anatomy can assume diverse patterns of epicardial distribution, they will all originate from the aortic sinuses facing the pulmonary trunk. Simple transpositions will show and isolated ventriculoarterial discordance which is seen in 50% of the cases. Complex transpositions include all cases along with either of the following [1].

Out of the above, the most common one is the ventricular septal defect, which can vary in location or size. The outlet and trabecular septum may show some degree of malalignment. There is usually a presence of left ventricular outflow tract obstruction in about 1/8th

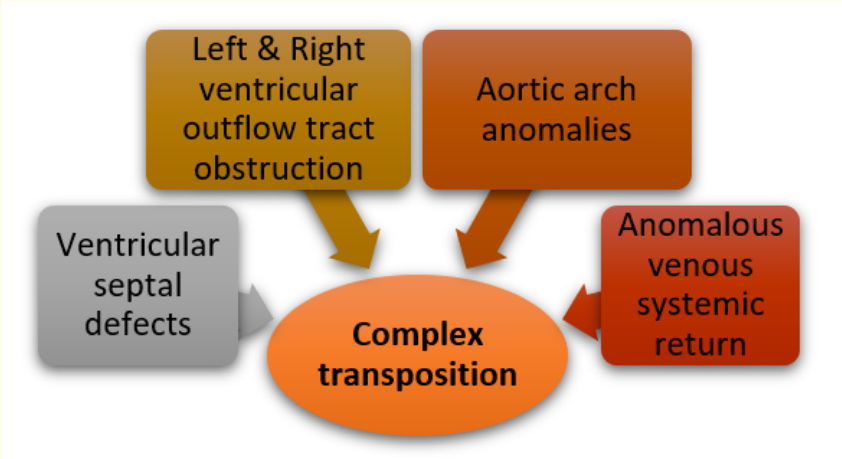

Figure 1

to 1/3rd of the cases affected with the presence of a ventricular septal defect. The obstruction of blood flow caused by this can give rise to a lot of valvular or subvalvular lesions.

The pulmonary and systemic circulations run in parallel as opposed to in series in the hearts affected by concordant atrioventricular and discordant ventriculoarterial connections.

The lungs and the left cardiac chambers (left atrium and ventricle) are responsible for dealing with the oxygenated blood. Whereas the systemic blood is flow begins and ends at the right cardiac chambers. Both these blood flows follow a closed circuit of circulation. And in such a pathological condition, only if there is acceptable mixing between both the circulations is survival possible. Though the exact etiology of the disease remains the following risk may result in the condition [5,6].

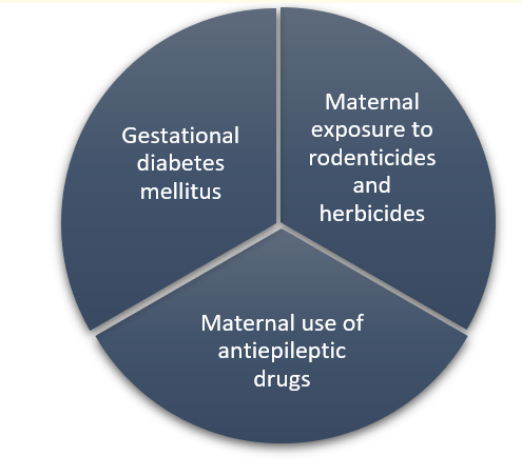

Figure 2

Clinical presentation

During infancy, the following features can be seen [1]:

- A severe hypoxemic state because of central cyanosis is seen.
- Bluish discoloration of the skin and mucosa.
- Progressive and profound central cyanosis seen within the first hours of life.
- Marked cyanotic state due to the reduced blood flow to the pulmonary vascular bed.
- The absence of an obstructive lesion with a large ventricular septal defect can allow the satisfactory mixing of both the circulations. In such cases, the cyanosis may not be present generally and is seen only during crying or agitation. This is because of the signs of congestive heart failure is more evident due to the excessive ventricular workload.
- Other conditions like tachypnoea, tachycardia, reduced weight gain, rhythm gallop, hepatomegaly, and diaphoresis are seen.
- Seldom heart murmurs, persistent arterial duct of the septal defect are heard though not constant [1].

Diagnosis

Careful history taking from the parents and thorough clinical examination must be done on the patient under good light conditions, and clinical manifestations must be properly assessed for. Definitive diagnosis relies on echocardiography, where the morphological and functional assessment can be done accurately. The transposition of the great arteries can be assessed by observing the specific features. The atrioventricular concordance can be assessed in a four-chamber view. However, he ventriculoarterial discordance is usually assessed otherwise. the vessel which arises from the left ventricle has a posterior course that bifurcates immediately which is identified as the pulmonary trunk in a five-chamber, parasternal long-axis, or even subcostal view [9].

The right ventricle is connected to the vessel that gives out the aorta. The short-axis view shows the pulmonary trunk in a central position where anteriorly and to the right, the aorta can be seen. It is important to assess the evaluation of the coronary artery pattern and to exclude other malformations prior to surgery [1,7].

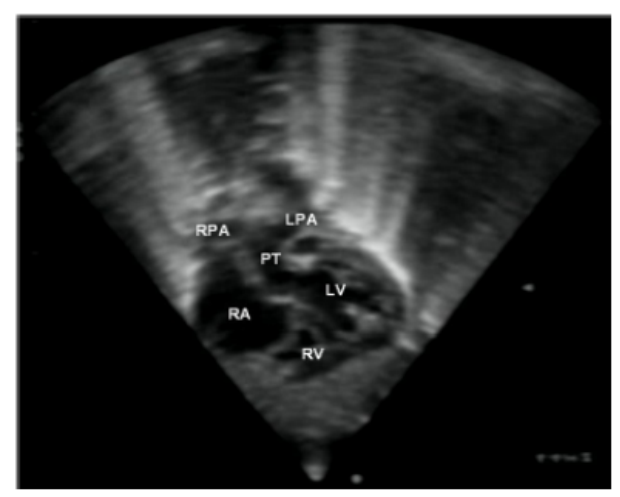

Figure 3a: *Subcostal view: showing a vessel originating from the left ventricle which bifurcates; identified as the pulmonary artery [1].*

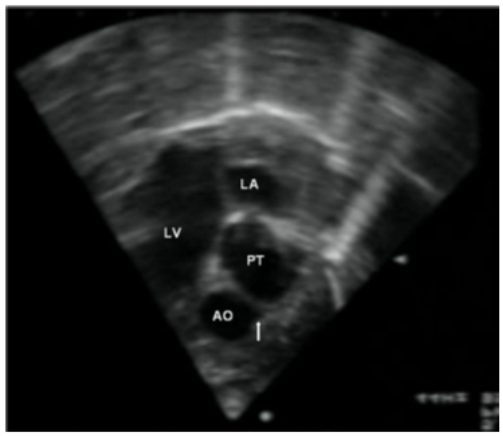

Figure 3b: *Short axis view: showing coronary arteries originating from the aorta in an anterior position and to the right [1].*

The two-dimensional echocardiography can be enhanced in its functional information by the doppler study. The indicator of adequate mixing between the systemic and pulmonary circulation is assessed after visualizing the flow between the arterial duct and septal defects. The measurement of gradients through the obstructive lesions and assessment of the function of atrioventricular and semilunar valves can also be done. The use of Computed tomography and magnetic resonance imaging can provide additional information if there is a presence of any associated lesions [8].

To reduce morbidity and mortality, it is best to opt for prenatal echocardiography. This can improve neonatal management. This method of prenatal echocardiography has proved to be very accurate and extremely helpful in planning appropriate surgery apart from the accurate diagnosis of the disease [9]. The differentiation between cyanosis due to cardiac disease or those that are caused by neurological or pulmonary disorders can be performed by a hypoxia text, which provides a simple means of assessing PaO_2 less than 50 mmHg [1].

Management

Palliative treatment

The preliminary aim to manage an affected newborn is to ensure the intercirculatory mixing of the blood. If the newborn presents with an atrial or ventricular septal defect, acceptable mixing is seen. This defect can be corrected at a later stage. However, this is rarely a common scenario, and in most cases, the first line of treatment is required.

The infusion of intravenous prostaglandin E1 can maintain the patency of the arterial duct, which can increase the pulmonary blood flow, which in turn increases the pulmonary venous return and left atrial pressure. This promotes left to right flow at an atrial level [10].

Satisfactory oxygenation of the systemic blood is seen with intercirculatory mixing and prostaglandin action in both simple and complex transposition. In 1966 the introduction of the Balloon atrial septostomy, also known as the Rashkind procedure, played an important role in the intervention of certain forms of congenital disease. This procedure can be used as a pre-operative management procedure in babies with transposition of the greater arteries [11].

A balloon-tipped catheter was placed in the left atrium through the oval foramen. The balloon, which is then inflated, is pulled back into the right atrium, which tears the atrial septum. The procedure is performed under the supervision of echocardiography, which provides

consistent visual guidance without the exposure of ionizing radiation in some centers. The risk of complications is minimized and allows access through the interatrial orifice [11].

The success of the procedure occurs only when the atrial septal defect has been reduced to at least 5 mm in diameter and where there is an increased flapping motion of the inferior rim of the atrial septum with an increase in the oxygen saturation. A long-lasting adequate interatrial communication can be created by balloon septostomy which is an effective and safe procedure. Aggressive surgical interventions like blade atrial septostomy and surgical atrial septectomy have been replaced by this procedure. In unstable newborns with severe hypoxemia, mechanical ventilation and oxygen are required to optimize the clinical condition. However, these measures may not universally benefit all patients. Excessive alveolar distension should be avoided by aggressive ventilator settings (Positive Inspiratory Pressure and Positive End Expiratory Pressure so that intercirculatory shunts are not disturbed. Bicarbonate should be sed to correct metabolic acidosis, as this can compromise the myocardial function. Inotropic agents or diuretics can be used in the case of cardiac failure [12].

Corrective treatment

The arterial switch operation (ASO) has become the preferred option for treating TGA because of its excellent outcomes due to the anatomical correction. Earlier, the application of ASO could not be made in newborns as early as required [4]. Balloon atrial septostomy (BAS) allowed infants to undergo surgery within the first month of birth. However, this procedure had complications such as atrial perforation, vascular injury, arrhythmia, and pericardial tamponade and increased the risks of cerebral embolism and stroke [2].

Nevvazhay., *et al*. in 2012 suggested that ASO must be performed as early as possible in infants with TGA without performing the BAS procedure [13]. In order to avoid the deterioration of left ventricular function, reduce postoperative complications and mortality, and thus effectively reduce hospital stay and costs it has been suggested to perform the ASO as early as possible. In this procedure, sectioning of the aorta and pulmonary trunks are done, and their distal extremities are transposed and anastomosed. The coronary arteries are then translocated to the neo-aorta.it is recommended to do this procedure in the first month of birth [14].

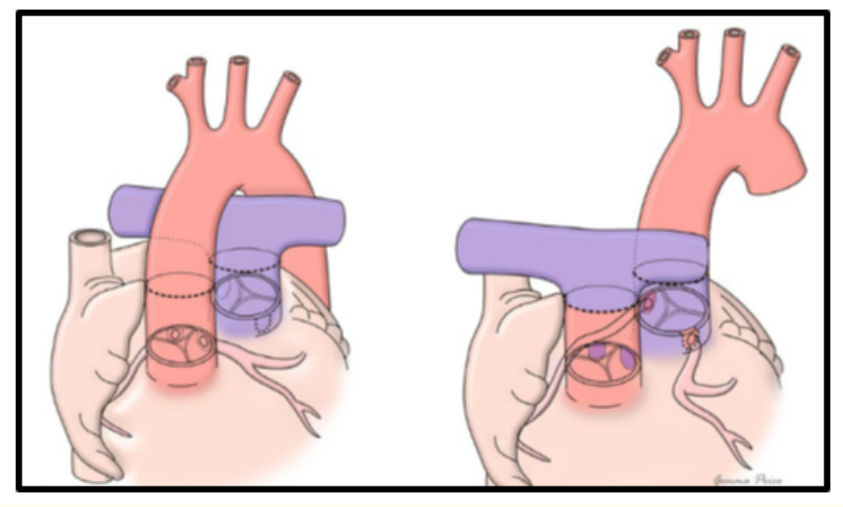

Figure 4: *Arterial switch procedure. Showing the connection of the proximal great arteries to the distal end of the other great artery, as well as the transfer of coronary arteries to the new aorta [1].*

Wrong or missed diagnosis and preoperative complications, such as multiorgan failure, renal failure, active infection, severe acidosis, or sub arachnoidian hemorrhage, can affect the timing of this procedure. If the palliative procedure has a poor prognostic outcome, it is better to switch to the surgical methods of treatment [14].

A newborn, if presents with a specific context of transposition with an intact septum, must be operated, preferably within the first two weeks of life. This ensures that the left ventricle does not have a significant involution, and its contractility can still support the systemic circulation. The left ventricle may need to be retrained in older neonates and young infants, before attempting the arterial switch, by pulmonary artery banding, (with or without an associated Blalock-Taussig shunt). The goal of this step is to establish a left/right ventricular pressure ratio between 0.6/0.75 without the pulmonary blood flow being compromised [14].

The arterial switch procedure must be tailored towards the individual morphological aspects in complex transposition. To repair concomitant malformations, complementary interventions may be necessarily associated with atrial or septostomy defect, can be easily closed by direct suture. A patch to close the communication of the ventricular septal defect but can be left open if it is very small. A parallel repair is required for an obstruction within the aortic arch, or the usage of a pulmonary homograft to enlarge the aorta if required [14].

If a surgical switch is not a feasible alternative procedure as shown below can be used [1].

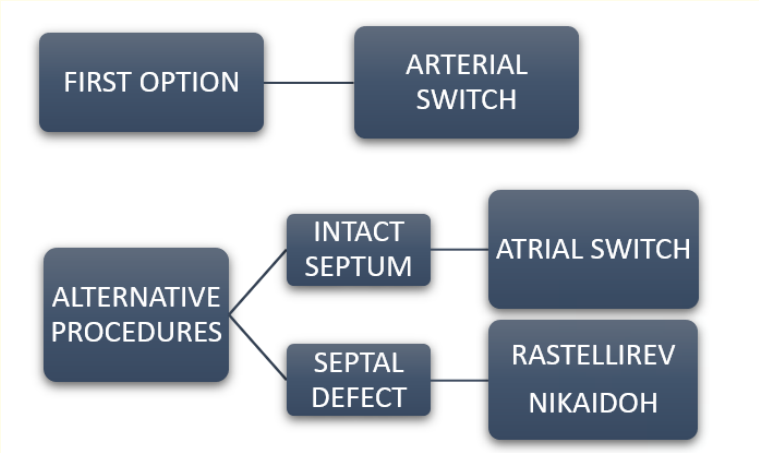

Figure 5: *Surgical procedures used in the correction of the transposition of the great arteries.*

Mustard or a Senning procedure can be done at an atrial level suitable for hearts with an intact ventricular septum.at the atrial level, the systemic venous return is redirected to the left ventricle and the pulmonary venous blood to the right ventricle. An extracardiac conduit is placed between the left ventricle and the pulmonary trunk where there is obstruction at the left ventricular outflow tract [1].

Complications seen with this procedure include:

- Sinus node dysfunction, obstruction to either pulmonary or systemic venous return,
- Supraventricular tachyarrhythmias, residual interatrial shunt,
- Right ventricular dysfunction and
- Pulmonary vascular obstructive disease.

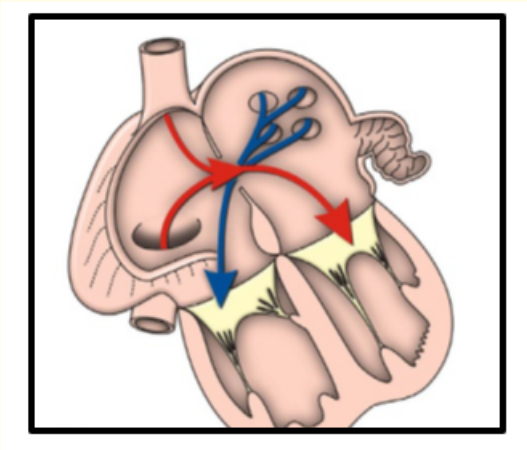

Figure 6A: *Figure showing redirection of pulmonary and systemic venous flows at atrial level [1].*

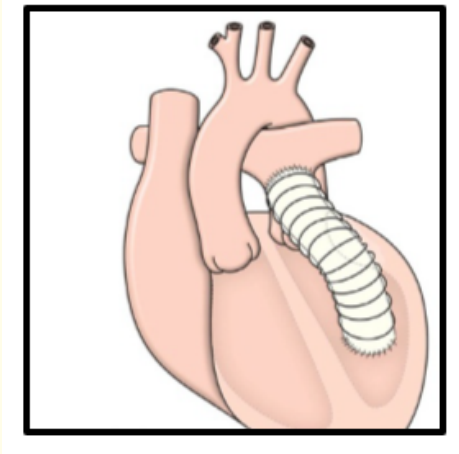

Figure 6B: *An external conduit (B) is placed to overcome the obstruction within the left ventricular outflow tract [1].*

The REV procedure or its modification and the Rastelli operation are commonly used in the ventricular septal defect. An intraventricular tunnel is passing through the septal defect in both the operations to create a connection to the left ventricle and the aorta. An extracardiac conduit is placed to connect the right ventricle to the pulmonary artery in the Rastelli procedure. The REV technique done through the LeCompte maneuver brings the pulmonary trunk forward, allowing its direct implantation in the right ventricle. Complications such as coronary ischemia, left ventricular dysfunction, other surgical complications like acute renal insufficiency must be anticipated with any surgical intervention [15].

Conclusion

TGA is a complex congenital life-threatening condition seen commonly in infants. There has been an increase in the fetal ultrasound scanning done to screen for congenital heart diseases in developed countries. The detection of specific congenital defects can have positive

consequences on the management of the fetus with the affected condition. The outcome of the diagnosis of the fetuses with heart conditions in utero can be affected by a number of conditions. Early detection of conditions like chromosol anomalies or heart conditions can lead to the early termination of pregnancy, which is an ethical concern as well. The treatment of transposition of arteries was restricted to palliative management until the middle of the 1920s. The idea about the history or prognosis of the disease was also limited. The advent of new and improvised surgical interventions and the improved quality of post-operative care has shown a change in the scenario where there is an increase in the survival rates of these patients. The improved methods show a marked improvement in cognitive functioning and an increased quality of life as well [8]. Infants diagnosed with TGA must undergo the procedure of ASO as early as possible. The left ventricular function should be comprehensively assessed in infants who have already passed the best treatment period. Satisfactory survival rates reduced reintervention, and lack of major complications during follow up makes it easier to perform the corrective surgeries at an earlier stage.

Improvement in terms of modalities of prenatal diagnosis and methods to minimize risk during surgical interventions may still be required to improve the quality and reduce morbidity and mortality of patients affected with TGA [8].

Bibliography

1. Martins P and Castela E. "Transposition of the great arteries". *Orphanet Journal of Rare Diseases* 3.1 (2008): 27.
2. Villafañe J., *et al*. "D-transposition of the great arteries: the current era of the arterial switch operation". *Journal of the American College of Cardiology* 64.5 (2014): 498-511.
3. Ho SY. "Color atlas of congenital heart disease". Mosby Limited (1995).
4. Wu QY., *et al*. "Surgical Treatment of Complete Transposition of the Great Arteries in Newborn". *Chinese Medical Journal* 129.19 (2016): 2381.
5. Becerra JE., *et al*. "Diabetes mellitus during pregnancy and the risks for specific birth defects: a population-based case-control study". *Pediatrics* 85.1 (1990): 1-9.
6. Abu-Sulaiman RM and Subaih B. "Congenital heart disease in infants of diabetic mothers: echocardiographic study". *Pediatric Cardiology* 25.2 (2004): 137-140.
7. Pasquini L., *et al*. "Coronary echocardiography in 406 patients with d-loop transposition of the great arteries". *Journal of the American College of Cardiology* 24.3 (1994): 763-768.
8. Bonnet D., *et al*. "Detection of transposition of the great arteries in fetuses reduces neonatal morbidity and mortality". *Circulation* 99.7 (1999): 916-918.
9. Pascal CJ., *et al*. "An echocardiographic study of diagnostic accuracy, prediction of surgical approach, and outcome for fetuses diagnosed with discordant ventriculo-arterial connections". *Cardiology in the Young* 17.5 (2007): 528-534.
10. Tálosi G., *et al*. "Prostaglandin E [1] treatment in patent ductus arteriosus dependent congenital heart defects". *Journal of Perinatal Medicine* 32.4 (2004): 368-374.
11. Lin AE., *et al*. "Balloon and blade atrial septostomy facilitated by two-dimensional echocardiography". *The American Journal of Cardiology* 57.4 (1986): 273-277.
12. Perry LW., *et al*. "Echocardiographically assisted balloon atrial septostomy". *Pediatrics* 70.3 (1982): 403-408.

13. Nevvazhay T., *et al*. "Arterial switch in the first hours of life: no need for Rashkind septostomy?" *European Journal of Cardio-Thoracic Surgery* 42.3 (2012): 520-523.

14. Planche C., *et al*. "Arterial switch". *Pediatric Cardiology* 19.4 (1998): 297-307.

15. Turon-Vinas A., *et al*. "Characteristics and outcomes of transposition of great arteries in the neonatal period". *Revista Española de Cardiología (English Edition)* 67.2 (2014): 114.

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