

Dextro-Transposition of the Great Arteries (D-TGA): Information

Page Links: [Definition](#), [Vascular Arrangement](#), [Circulation](#), [Standardized Definition](#), [Incidence](#), [Associated Cardiac Malformations](#), [Extracardiac Malformations](#), [Chromosomal Abnormalities](#), [Treatment](#), [Surgical Management](#), [References](#).

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Definition

The primary arteries or great vessels are the aorta and pulmonary artery, which originate from the left ventricle and right ventricle, respectively.

In dextro-transposition of the great arteries (D-TGA), the aorta arises from the right ventricle instead of the left ventricle, and the pulmonary artery arises from the left ventricle instead of the right ventricle. This arrangement is termed **ventriculoarterial discordance**.

In transposition of the great arteries (TGA), the right atrium normally connects to the right ventricle via the tricuspid valve while the left atrium normally connects to the left ventricle through the mitral valve. This arrangement is termed **atrioventricular concordance**.

Vascular Arrangement

In D-TGA, the **pulmonary artery** is the **posterior artery** and branches after arising from the left ventricle. The pulmonary artery appears as a straight coursing artery when arising from the left ventricle and the lateral branching helps to define it. [\[1\]](#)

The **aorta** is the **anterior artery** arising from the right ventricle. The head neck vessels come off from the aorta.

Circulation

After birth, D-TGA is compatible with life only if the ductus arteriosus and/or the foramen ovale remain patent to permit mixing of oxygenated blood. Otherwise, there are two separate parallel circulations. The aorta from the right atrium and right ventricle carries deoxygenated blood to and from the systemic circulation, while the pulmonary artery from the left atrium and left ventricle carries oxygenated blood to and from the lungs. The deoxygenated blood flowing to the cerebral circulation and heart results in hypoxia and neonatal cyanosis, and neonatal death occurs if emergent management is not undertaken.

Standardized Definition

To standardize the nomenclature for D-TGA, the following categories are suggested: TGA with intact ventricular septum, TGA with ventricular septal defect (VSD), and TGA with VSD and left ventricular outflow tract obstruction (LVOTO). [\[2\]](#)

Incidence

The incidence of D-TGA is 0.32 per 1000 live births. [\[3\]](#) D-TGA is a relatively frequent anomaly among CHD patients, comprising 5.7% of that group in one study. [\[4\]](#) The male to female ratio in D-TGA is greater than 2:1 in a number of studies. [\[5\]](#)

Associated Cardiac Malformations

A ventricular septal defect (VSD) is the most commonly associated defect in patients with D-TGA. In a review of 261 D-TGA cases, the following distribution of

associated defects was noted [\[6\]](#):

1. Any associated cardiac defect: 22%
2. VSD only: 15%
3. VSD plus coarctation of the aorta: 6.5%
4. Coarctation of the aorta only: 0.3%

In another series of D-TGA, isolated VSD occurred in 7 of 18 cases. VSD was also associated with pulmonary stenosis, atrioventricular septal defect, and one case each of hypoplastic left heart and tricuspid atresia. [\[7\]](#)

D-TGA is also associated with abnormal course of the umbilical vein, absent ductus venosus [\[8\]](#), and right atrial isomerism. [\[9\]](#)

Extracardiac Malformations

The prevalence for extracardiac malformations is considered low in D-TGA. [\[10\]](#) In one study of 124 cases of D-TGA, only eight (6.5%) demonstrated abnormalities that were detected on prenatal ultrasound. [\[11\]](#) In a smaller study, 6 of 18 patients demonstrated extracardiac anomalies, but no extracardiac malformations were noted among corrected D-TGA patients. [\[12\]](#)

Chromosomal Abnormalities

The prevalence for chromosomal abnormalities is low in D-TGA. [\[13\]](#) No chromosomal abnormalities were noted in 17 of 18 cases of D-TGA. [\[14\]](#)

Treatment

After birth, a continuous infusion of prostaglandin E1 (PGE1) is needed to maintain ductus arteriosus patency allowing a mixture of oxygenated blood to perfuse the myocardium and brain. If there is significant hypoxia due to inadequate mixing of

the oxygenated and deoxygenated blood, a balloon atrial septostomy is performed to enlarge the opening in the atrial septum, which may be life saving. [\[15\]](#)

Surgical Management

The surgical procedure of choice for D-TGA is the arterial switch operation. [\[16\]](#) This procedure restores the correct anatomic relationship between the pulmonary artery and aorta with the right and left ventricle respectively. In centers with surgical expertise, survival probability is reported above 90% at 10 years in an unselected population. [\[17\]](#) Various coronary artery distribution patterns are possible. A complex and challenging part of the procedure involves the excision and relocation of the coronary arteries from the native aortic root to the neo-aortic root.

References

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