

CASE REPORT: IN UTERO ECHOCARDIOGRAPHIC DIAGNOSIS OF CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES WITH ASSOCIATED DEXTROCARDIA

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ABSTRACT

Congenitally corrected transposition of the great arteries (cTGA), defined by discordant atrioventricular and ventriculoarterial connections, is an uncommon congenital cardiac malformation. Although hemodynamic consequences should not arise, in theory, because these misconnections cancel each other. But it rarely exists without further associated cardiac anomalies that worsened the prognosis. We describe a fetus that was diagnosed echocardiographically with cTGA and dextrocardia, without additional cardiac anomalies, in utero at 24 weeks and 3 days's gestation. Actually, the patient is still pregnant.

Key words: *Fetal echocardiograph; Dextrocardia; Corrected transposition of the great arteries*

RIASSUNTO

La trasposizione corretta delle grandi arterie (cTGA), caratterizzata da discordanti connessioni atrio-ventricolari e ventricolo-arteriose, è una rara malformazione cardiaca congenita. Nonostante ciò le conseguenze emodinamiche, in teoria, non dovrebbero sorgere, in quanto queste alterazioni si dovrebbero annullare a vicenda. Tuttavia tale alterazione raramente esiste senza ulteriori anomalie cardiache associate, le quali peggiorano la prognosi. Descriviamo un feto a 24 settimane e 3 giorni di gestazione al quale è stata diagnosticata una cTGA con dextrocardia tramite ecocardiografia, senza ulteriori anomalie cardiache associate. La paziente, attualmente, è ancora in stato di gravidanza.

Parole chiave: *Ecocardiografia fetale; Dextrocardia; Corretta trasposizione grandi arterie*

INTRODUCTION

Congenitally corrected transposition of the great arteries (cTGA) is rare cardiac defect, occurring in approximately 0.7 % of all live births with congenital heart disease.

It is characterized by discordant atrioventricular and ventriculoarterial connections because the atria connecting with anatomically discordant ventricles and the ventricles connecting with discordant and transposed great arteries.

This double discordance results in a physiologically hemodynamic compensation, but the left ventricle supports the pulmonary circulation, hence there is a normal arterial oxygen content, and the right ventricle supports the systemic circulation.

However, isolated forms of cTGA rarely exists and they are associated with other major cardiac anomalies, the most common of which are ventricular septal defect, pulmonary outflow obstruction, tricuspid valve (systemic) Ebstein-like deformity, and rhythm disturbances that can influence their prognosis.¹

MATERIALS AND METHODS

In this report, a 38-year-old woman was referred to our Center at 24 weeks and 3 days' gestation because of suspected fetal cardiac anomalies. There was no familial history of congenital heart disease, and two previous spontaneous abortions had been recorded. Patient had no known systemic disease. She didn't perform amniocentesis. Morphologic scan of fetus one week before evaluated a normal intrauterine fetal growth.

A Voluson 730 ultrasound apparatus was used for fetal echocardiography in which dextrocardia, visceral situs solitus, viscerotrial concordance, and atrioventricular and ventriculoarterial discordance were found.

We diagnose heterotaxy or situs ambiguus if some of the organs are on the correct side while others are on the opposite of their expected side. Heterotaxy can be, either, divided according to which organ is

abnormal, into situs solitus with dextrocardia, as in this case reported, in which the position of the major cardiac axis (from the base to the apex along the interventricular septum) points to the right instead the stomach remains on the left side, and visceral situs on the contrary. (fig. 1)

A detailed examination of the ventricular morphology helped to confirm that the right-sided right atrium entered, via a morphologic mitral valve (MV), the right-sided left ventricle (LV), which in turn emptied into the posterior, right-sided pulmonary artery instead the left-sided left atrium was connected, through a morphologic tricuspid valve (TV), to the left-sided right ventricle (RV), which emptied into the anterior, left-sided aorta. (fig 2-3-4)

The MV/TV and LV/RV ratios were above the normal range.

The ascending, descending aorta and the aortic isthmus were normal in size. (fig. 5)

Actually, the patient is still pregnant.

Fig 1. Situs solitus with dextrocardia . The heart is on the right side, opposite of stomach that remains on the left.



Fig. 2. Note the ventriculoarterial discordance: the aorta (AO) arises from the left-sided, morphologically right ventricle instead bifurcating pulmonary artery (POLM) arising from the morphological left ventricle



Fig. 3. ColorDoppler view: bifurcating pulmonary artery arises from the morphological left ventricle.

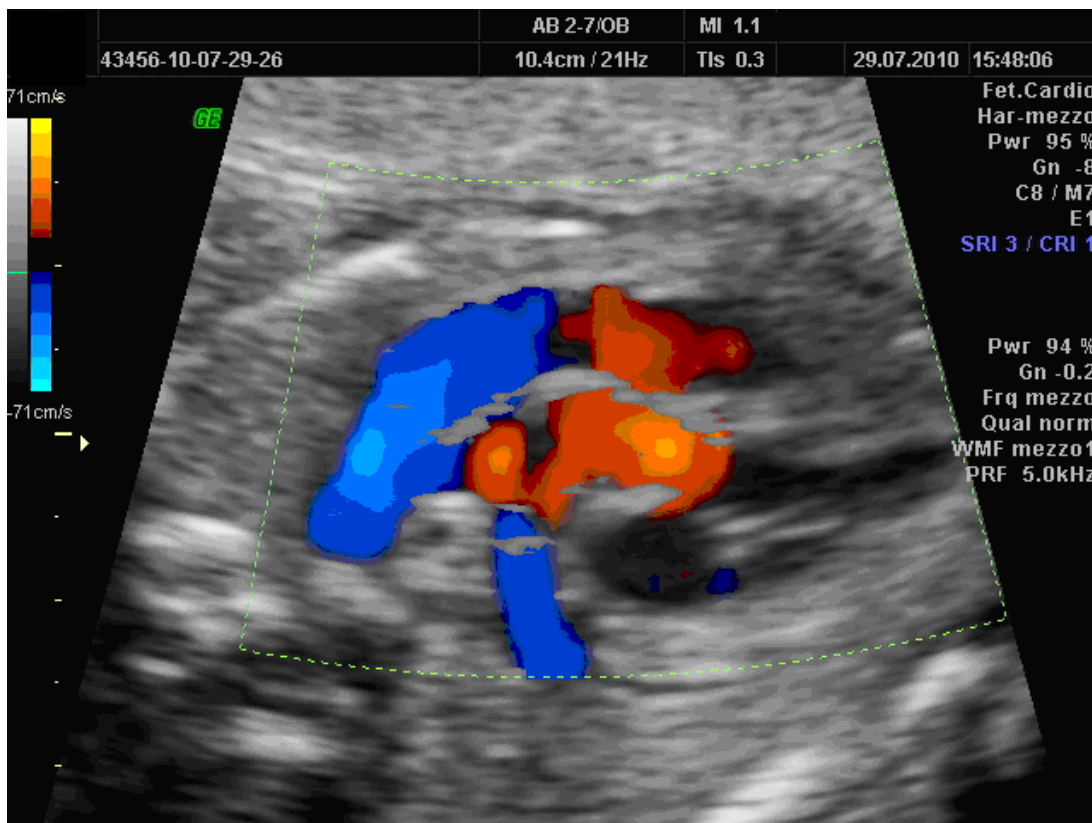


Fig 4. The 3-vessels view. The pulmonary bifurcating (1-2) is in the middle between ascending aorta and superior vena cava, almost aligned.



Fig 5. Sagittal view of the fetal heart demonstrates an aortic arch connecting to the right ventricular outflow tract.



DISCUSSION

Despite hemodynamic compensation, the long-term prognosis of ccTGA is unclear because of the possible development of heart failure later in life originating from right ventricular dysfunction, tricuspid regurgitation or conduction abnormalities till complete atrioventricular block.²⁻³⁻⁴

CONCLUSION

In conclusion, we think that a fetal accurate segmental analysis of the cardiac chambers and great vessel connections and relations by fetal echocardiography could be highly helpful in improving follow-up of potentially critical neonates even when they are initially asymptomatic and in counselling of parents prenatally.

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