Truncus arteriosus – echocardiographic findings
Truncus arteriosus – achados Ecográficos

Sofia Mendes*, Cláudia Araújo**, Rui Carvalho***
Hospital de Santa Maria Centro Hospitalar de Lisboa Norte

Abstract

Truncus Arteriosus (TA) is a rare congenital cardiac malformation in which a single common artery arises from the heart and supplies the systemic, pulmonary, and coronary circulations. Associated findings are common and include a truncal valve and a ventricular septal defect. Noncardiac congenital malformations may also be present.

Prenatal diagnosis can be challenging, since differential diagnosis with tetralogy of Fallot and pulmonary atresia is sometimes difficult.

Keywords: Truncus arteriosus; Prenatal diagnosis

Truncus arteriosus (TA) is a rare congenital cardiac malformation, with a reported incidence of 6-10 cases per 100000 live births[1,2].

It is characterized by a single arterial vessel originated from the heart, which overrides the ventricular septum and supplies the systemic, pulmonary and coronary circulations. Other associated findings are common and include ventricular septal defect and aortic arch anomalies. Noncardiac congenital malformations may also be present[3].

Truncus arteriosus has been classically classified into four types depending on the site of origin of the pulmonary arteries[4]. In type I a short pulmonary trunk gives rise to the pulmonary arteries. In type II and type III there is no main pulmonary artery, but in the former the branch pulmonary arteries arise from posterior aspect of trunk close to each other just above the truncal valve and in the later both branch pulmonary arteries arise from either side of the truncus, separate from each other. In type IV the truncus is associated with an interrupted aortic arch.

Pre-natal diagnosis can be challenging, since differential diagnosis with tetralogy of Fallot and pulmonary atresia is sometimes difficult[5]. Causes of TA are not entirely known, but it seems to be associated with a microdelection within chromosome 22[6]. Similar to other conotruncal anomalies TA is not associated with alteration of fetal hemodynamics. However, after delivery, with loss of physiologic shunts, pulmonary congestion and heart failure develop. If left untreated, TA is associated with a high mortality rate within the first year of life.

The images concerns to a pregnancy in which, because of a positive first trimester screening test, a fetal chromosomal analyses was offered and a partial duplication of chromosome 22q was detected. At the second trimester scan fetal micrognatia, nuchal oedema and absent nasal bone were detected and fetal cardiac examination revealed normal apical four –chamber view. However when moving the transducer cranially in order to visualize cardiac outflow tracts, a single arterial trunk arising from the ventricules overriding the ventricular septum was seen (Figure 1). A subtruncal ventricular septal defect was also present. Pulmonary arteries originated form a short pulmonary trunk arising from the truncus. On color Doppler imaging, blood flow from both ventricules could be seen entering the single outflow tract (Figure 2) and there was evidence

*Interna do Internato Complementar de Obstetrícia e Ginecologia CHLN/HSM
**Assistente Hospitalar de Obstetrícia e Ginecologia, Unidade de Ecografia do Departamento de Obstetrícia e Ginecologia e Medicina da Reprodução do CHLN/HSM
***Assistente Hospitalar de Obstetrícia e Ginecologia, Unidade de Ecografia do Departamento de Obstetrícia e Ginecologia e Medicina da Reprodução do CHLN/HSM. Assistente Convidado da Faculdade de Medicina da Universidade de Lisboa – Clínica Universitária de Obstetrícia e Ginecologia

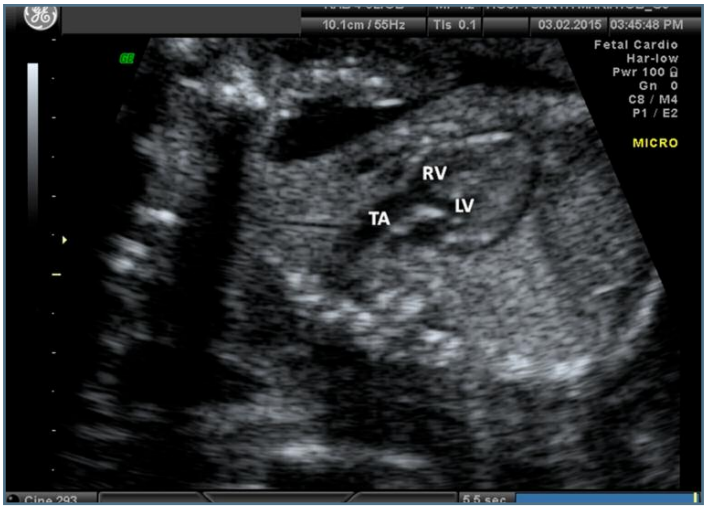

FIGURE 1. Single large outflow tract equally overriding the ventricular septum
LV – Left ventricule; RV – Right ventricule; TA – Truncus arteriosus

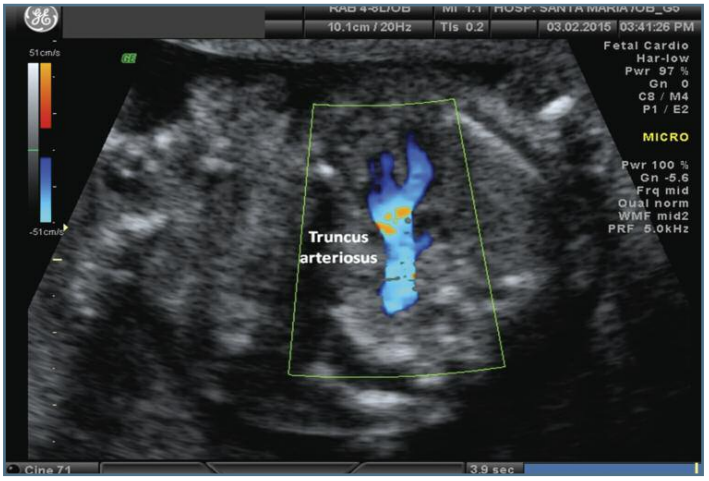

FIGURE 2. Color Doppler imaging – blood flow from both ventricles entering the single outflow tract

of mild regurgitation across a truncal valve. These findings are highly suggestive of truncus arteriosus type I.

REFERENCES

1. Reller MD, Strickland MJ, Riehle-Colarusso T, Mahle WT, Correa A. Prevalence of congenital heart defects in metropolitan Atlanta, 1998-2005. J Pediatr 2008;153(6):807.

2. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol 2002;39(12):1890.

3. Radford DJ, Perkins L, Lachman R, Thong YH, Spectrum of Di George syndrome in patients with truncus arteriosus: expanded Di George syndrome. Pediatr Cardiol 1988;9(2):95-101

4. Collet RW, Edwards JE. Persistent truncus arteriosus; a classification according to anatomic types. Surg Clin North Am 1949; 29(4):1245.

5. Swanson TM, Selamet Tierney ES, Tworetzky W, Pigula F, McElhinney. Truncus arteriosus: diagnostic accuracy, outcomes, and impact of prenatal diagnosis. Pediatr Cardiol 2009;30(3):256-61.

6. Momma K, Cardiovascular anomalies associated with chromosome 22q11.2 deletion syndrome. Am J Cardiol 2010;105(11): 1617.