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Journal of Clinical and Experimental Cardiology

Editorial

A rare association of double outlet right ventricle with non-committed interventricular communication, aberrant right subclavian artery, persistent left superior vena cava and tracheoesophageal fistula in a newborn

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ABSTRACT

Double Outlet Right Ventricle (DORV) is associated to a wide set of cardiovascular anomalies. We report a rare combination of congenital cardiac malformation of DORV with unrestrictive non-committed interventricular communication, persistent left superior vena cava, sub pulmonary stenosis and aberrant right Subclavian artery.

Keywords: Congenital heart disease; Double outlet right ventricle; Left superior vena cava; Aberrant right subclavian artery;

EDITORIAL NOTE

The persistence of Left Superior Vena Cava (LSVC) is the most common anomaly of the systemic venous returns [1]. It's often an incidental finding and the prevalence is < 0.5% in general population. The diagnosis of persistent LSVC may be isolated or in association of Congenital Heart Disease (CHD), where the incidence can reach 4% [2].

Ari et al. reported data of 88 patients affected by CHD and persistent LSCV, among them only 14.6% was associated to Double Outlet Right Ventricle (DORV).

We would like to report to your attention our singular case of a 2-day-old newborn, of body weight 3,080 Kg with postnatal diagnosis of LSVC and DORV with a set of rare anomalies. Echocardiogram at birth showed DORV (S, D, D), LSVC draining in a dilated coronary sinus without bridging vein, unrestrictive non-committed InterVentricular Communication (IVC), located in the inlet septum, bilateral infundibulums, subpulmonary stenosis, left sided normal aortic arch and a suspected Aberrant Right Subclavian Artery (ARSA). CT scan angiography confirmed diagnosis and showed the presence of tracheoesophageal fistula "H" shape with a valve mechanism. Not chromosomal anomalies were detected. DORV is a complex CHD with various common and rare subtypes as DORV with non-committed ventricular septal defect which is reported in 10%-20% of cases [3].

Freedom described a wide range of associate anomalies occurring in DORV [4].

In our case we defined the hole between the ventricles as IVC in order to anatomical and morphological description given by Anderson, who defined IVC in DORV as the space between the ventricles on the basis of the continuation of the long axis of the muscular septum, in preference to VSD, because the outlet septum is of necessity exclusively right ventricular when both arterial trunks arise from the RV [5-7].

Ramaswamy et al. reported that generally, considering all CHDs, ARSA is more frequently discovered in tetralogy of Fallot and in their report among 226 cases of ARSA, only 5 patients presented a DORV associated [8]. Zapata and colleagues analyzed more of four thousands of CHD and discovered the presence of ARSA only in 117 cases; considering only patients with ARSA and CHD, 38% were conotruncal anomalies, of which 11 of 117 patients were affected by DORV [9]. Moreover, the association of ARSA and tracheoesophageal fistulas/esophageal atresia is rarely reported, about 12% [10].

The attempts to classify the DORV are different [11-13] but the major problem is always represented by the surgical challenging indication. The purpose of this letter inspired by your article is to make readers understand that DORV is a complex disease that could be associated to much rare pathology. Our case is represented by a combination of uncommon cardiovascular and extra cardiac pathologies that make clinical and diagnostic

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Received: March 16, 2021; **Accepted:** March 30, 2021; **Published:** April 06, 2021

Citation: Caruso E, Farruggio S (2021) A Rare Association of Double Outlet Right Ventricle with Non-Committed Interventricular Communication, Aberrant Right Subclavian Artery, Persistent Left Superior Vena Cava and Tracheoesophageal Fistula in a Newborn. J Clin Exp Cardiolog.12: e672

assessment even more difficult. Management of these patients depend on extremely variable morphology and hemodynamic and echography and CT scan plays a crucial role in in defining the anatomy and morphology of DORV with associated anomalies.

AUTHORS CONTRIBUTIONS

Silvia Farruggio: wrote article and data interpretation.

Elio Caruso: wrote article, critical revision and approval of article.

CONFLICT OF INTEREST

None.

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