Cor triatriatum dextrum. Surgical treatment in a neonate

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This is the report of a cor triatriatum dextrum, detected in utero and surgically treated at 10 days of age. We have been unable to find a similar report of neonatal correction in the literature. (Acta Cardiol 2003; 58(1): 39-40)

Keywords: cor triatriatum dextrum – surgical neonatal correction.

Case report

A term baby girl was delivered after echographic in utero detection of right ventricular hypoplasia. Her birth weight was 3340 g and her clinical examination unremarkable except for an enlarged liver (2 cm below coastal margin) and cyanosis when crying. There was a soft diastolic rumble in the xyphoid area. At echography, there was an abnormal partition of the right atrium by a weblike membrane, between the patent foramen ovale (FO) and the tricuspid valve, suggesting the diagnosis of cor triatriatum dextrum (CTD), confirmed by angiography. Early surgery was recommended. At operation, the RA was enlarged contrasting with a very small RV. Through a right atriotomy, the large multiperforated web joining the superior and anterior caval ostia, was resected, the FO left patent. The baby rapidly did well and at 12 months of age, there was a normal growth curve and the girl had a normal echocardiography.

Discussion

CTD is a rare congenital heart disease, even less frequent than cor triatriatum sinistrum, a well-known anomaly. Embryologically, in early stages of cardiac development, the opening of the sinus venosus into the RA is guarded by two prominent folds, the right and left valves of the sinus venosus, which join superiority to form the septum spurium. As the sinus venosus becomes progressively incorporated into the RA, these valves regress. The remnant of the left venous valve fuses with the emerging atrial septum secundum to form the posterior portion of the atrial septum. At 12 weeks gestation, the right venous valve has normally disappeared, the crista terminalis is left as remnant of the superior portion and the Eustachian and Thebesian valves as remnants of the caudal portion at the lateral margins of the IVC and coronary sinus, respectively. During the remainder gestation, the Eustachian valve orients oxygen-rich blood from the IVC across the FO. The normal Eustachian valve is a weblike structure just anterior to the orifice of the IVC, seldom wider than 1 cm, with no known functional significance in extra-uterine life. If the right valve fails to regress, varying degrees of septation of the RA may result. Physiopathologically the RV and tricuspid valve are usually underdeveloped, due to low blood inflow. The condition is usually asymptomatic and diagnosed incidentally in late childhood or adulthood. It has even been discovered fortuitously at necropsy only. In the absence of communication between the atria, the symptoms are seldom prominent and resemble those of tricuspid stenosis or Budd-Chiari syndrome, which could even lead to fatal cirrhotic hepatic failure. Associated disorders include Ebstein’s anomaly, atresia of the tricuspid valve, atresia or stenosis of the pulmonary valve and anomaly of the pulmonary venous return. The best diagnosis mean is echography. Early surgery offers an opportunity for cure of this potentially lethal condition. Leaving the FO patent preserves the systemic cardiac output at the cost of a
transient desaturation awaiting spontaneous correction of the altered RV compliance and elevated perinatal pulmonary vascular resistances. Percutaneous balloon correction is a palliative option.

Acknowledgment

Thanks to Miss Renia-Laure for technical support.

Bibliography


Fig. 1. – During ventricular repletion, the abnormal membrane (AM) is quite separated from the tricuspid valves (TV). Intra-operative transoesophageal echography depicting the abnormal membrane with hypoplastic right ventricle.