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Case Report

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Unusual pulmonary venous return obstruction associated with atrio-ventricular septal defect

Abstract

Venous pulmonary return obstruction associated with atrio-ventricular septal defect (AVSD) has been already described. The anatomic substratum of this obstruction may vary from a patient to another. We report the case of a 20-month old infant who presented with signs of congestive right heart failure. The diagnosis of ASVD associated with unusual form of pulmonary venous return obstruction due to deviated interatrial septum was confirmed by imaging modalities. Treatment consisted on surgical repair of the AVSD and the obstruction removal to allow the restitution of the left ventricular volume.

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Introduction

Atrioventricular septal defect (AVSD) is a congenital heart disease often well tolerated during the first years of life in absence of associated important mitral valve regurgitation. We report the case of a child with a poorly tolerated AVSD due to pulmonary venous return obstruction caused by deviated intratrial septum.

Case report

We report a case of 20 months old baby girl admitted to our départements of pediatic cardiology for shortness of breath and frequent respiratory infections. At admission physical examination showed a decreased feeding , a poor weight gain (the height was 75 centimeters, weight was 7 kilograms and lower than the 3rd percentile).The baby presented a rapid breathing, Her blood pressure was 125/60 mmHg, her pulse rate was 85 beats/min, auscultation areas showed a grade 3/6 murmur audible on the left parasternal border, intensive murmur of tricuspid regurgitation and split S2 at the pulmonary valve. otherwhise She had right heart failure signs with spontaneous jugular turgesence, The abdominojugular reflux and peripheral oedema.

Electrocardiogram showed a sinus rythm, a QRS axis at –30° and rsR aspect' in lead V1. Chest radiography revealed a cardiothoracic ratio of 0.6, with dilated pulmonary trunk and increased pulmonary vasulature. Echocardiography showed a restrictive small primum atrioseptal defect (ASD) of 4 mm of diameter, a small restrictive ventricular septal defect (VSD) and two separate atrioventrular valves (AVVs) with a cleft in the anterior leaflet of the left-sided AVV (mitral valve). The particularity of echocardiograohic finding was a left deviation of of interatrial septum leading to malignement of left of atrioventricular valve us a consequence the venous pulmonary flow had to cross the restrictive ASD to reach the level of the left atrioventricular orifice figure. Concerning veins retrun we objectivated persistent left superior vena cava draining into the coronary sinus and normal connected pulmonary veins

The right ventricle was dilated, hypertrophied and clearly dominant. The left ventricle was smaller with dimensions at the lower limit of normal for the body surface, with the diastolic and telesystolic diameters measured at 34 mm and 15 mm, respectively. The atrioventricular annulus diameters measured respectively 15 mm to 14.5 mm on the left and 18.5 mm on the right, with an atrioventricular valve index (AVVI) of 0.78. Doppler showed an accelerated flow through the ASD with maximum velocity of 2 m/s, a mild mitral regurgitation, important, tricuspide regurgitation with a severe pulmonary arterial hypertension estimated at 75mmhg. We concluded to The diagnosis of AVSD associated to of venous pulmoary return obstruction secondary to deviated interatrial septum defect and restrictive interatrial septal defect reponsable to severe pulmonary hypertension. Patient was refered to surgical repair of ASV with excision of the venous obstacles to outflow The chest was explored by median sternotomy intraoperatively. Per operative exploration confimed echocardiographic finding, it was a posterior deviated septum directed towards venous pulmonary direction associated to a smalll interatrial septal defect type primiun causing obstacle to pulmonary vein return. this deviation mimied a supramitral membrane.

Te surgical procedure consisted of resection of the lower part (juxta valvular) of the interatrial septum and realign it to the inter ventricular septum , a closure of the ostium primum and finally a repair of the cleft of atrioventricule valve. The baby tolerated this procedure well. But few days later she developed a severe sepsis responsable for her death.[1,2]

Discussion

The association of atrioventricular septal defect AVSD with an obstruction of pulmonary venous return has been reported in the literature. In a multiinstitutional study. Published by Kort[3] in 2004, 13.7% of patients with pulmonary venous stenosis had AVSD and 0.9% of patients with AVSD had pulmonary venous return obstruction. The originality of our work consist in the nature of anatomic substrum of the pulmonary venous obstacles, that appear to be a malignement of the septum associated to restrictive. Such us aspect was not reported previously in fact in published reports[4–6] a supramitral mitral, a Cor triatriatum, or primitives anomalies of venus pulmonary valve. seemed to be the usual type of pulmonary venous return obstacle. Woong-Han[7] reported à case of malignement of interatrial septum associated to complete ASVD. This obstruction dont cause hemodynamic distrubance since the ASD was large allowing a normal flow across the mitral valve. Pardoxally in our case the AVSD was poorly tolerated because of the small diameter and the restrictive type of ASD.

For surgery therapy our stategy was simliar to that reported by Woong-Han[7] who opted for the same surgical technique. IN our patient, the indication of a biventricular repair was retained despite the clearly dominant aspect of the right ventricle, and was justified because baby has a favorable atrioventricular index (0.78) And especially based on the fact that the preload of the left ventricle was significantly lower comparing to that right ventricle due to the pulmonary venous obstacle and these anomalies will be reversible after releasing of obstruction. To date there are no formal recommendations to guide patients with poorly balanced ventricles to a single or biventricular treatment. Cohen[2] proposed an algorithm summarizing the therapeuttic strategy in management of ASVD, the choice of two ou single ventricular repair depends on the AVVI, the présence of ductal dependent circulation and the VSD finding. In our opinion, we must take into account the maximum parameters and especially analyze each case separately, considering both the morphological data (diameters and ventricular volumes, measurement of atrioventricular orifices) that hemodynamic data (ventricular load conditions).

Conclusion

Pulmonary venous return obstruction associated to AVSD is most commonly due to supra-mitral membrane, triatrial heart, or stenosis of pulmonary veins. The originality of this case lies in the fact that it illustrates an unsual and particular form of venous pulmonary obstruction at type of malignement of the septum with a restrictive ASD. The surgical cure is simple. The removal of the obstacle allows restitution of the left ventricular volume, often underestimated preoperatively because of its low preload.

Acknowledgments

None.

Conflicts of interest

Author declares that there is no conflicts of interest.

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