Uhl's anomaly associated with pulmonary atresia intact ventricular septum: Report of two cases.


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Introduction

Uhl's anomaly is rarely encountered anomaly. Absence of right ventricular myocardium may be the result of primary non-development of myocytes or may be due to selective apoptosis. Uhl considered the disease to be congenital in origin, since then, there have been numerous similar case reports of apparently congenital hypoplasia of the entire or near-entire right ventricle, with or without other associated congenital heart defects.

Case 1

The premature neonate was born at 35 4/7 weeks gestation and weighed 1510 g. His initial oxygen saturation was 65%. Echocardiography revealed enlargement of right atrium and ventricle, severe tricuspid insufficiency, and thin musculature of right ventricle and intact ventricular septum. Pulmonary antegrade blood flow could not be detected. It was provided by ductus arteriosus (Figure 1). The condition of the infant continued to deteriorate. Recurrent hypotension and severe cyanosis required volume and dopamine, prostaglandin infusions. The patient died at the two hours of life due to intractable heart failure.

Case 2

The second term neonate was born 3120 g. with oxygen saturation of 75%. Echocardiography revealed intact ventricular septum with pulmonary atresia associated with enlargement of right atrium and ventricle, severe tricuspid insufficiency with 2.2 m/sec velocity, and thin musculature of right ventricle. Angiography was performed for ductal stenting but unfortunately could not be performed due to ductal position.

Conclusion

Dysplasia of tricuspid valve, pulmonary atresia with intact ventricular septum have been described in association with Uhl's anomaly. The necropsy findings of a case of Uhl's anomaly associated with pulmonary atresia in a newborn was reported in the literature as an important reduction in the number of muscular fibers and a very thin right ventricular wall as described by Uhl. The prognosis of the complete form of Uhl's anomaly associated with pulmonary atresia with intact ventricular septum is poor as in these cases.

Figure 1: Echocardiographic examination of patient. Massive right ventricular and atrial dilatation, thin muscular layer of right ventricle. (RV; right ventricle, RA; right atrium)