SUCCESSFUL REPAIR OF COARCTATION OF THE AORTA AND OBSTRICTIVE TOTAL ANOMALOUS PULMONARY VEINS CONNECTION IN A ONE-DAY-OLD NEWBORN

Abstract

A clinical case of successful repair of a combination of total anomalous pulmonary venous connection (TAPVC) with collector stenosis in one-day-old newborn. Only one case of successful correction of such a pathology is reported previously. The operation was performed with cardiopulmonary bypass and temporary antegrade brain perfusion. The narrowed aortic area was resected, the integrity of the aortic arch was restored using an extended anastomosis, and TAPVC correction was performed using a “sutureless technique”. The postoperative period was uneventful. The newborn was discharged from hospital on the 12th day.

Keywords

newborn • coarctation of aorta • TAPVD

Introduction

Coarctation of the aorta makes up 6–8% of all congenital heart defects and is defined as a narrowing of the aortic lumen, sometimes associated with important intracardiac lesions. Total anomalous pulmonary venous connection (TAPVC) is a rare pathology and occurs in about 0.4% to 2% of congenital heart defects. Combinations of these pathologies are very rare.

Clinical Summary

The 2.9 kg newborn was admitted to the hospital on the first day of life in an extremely grave condition due to respiratory failure, coarctation of the aorta and supracardiac TAPVC with the proximal collector stenosis up to 65-70%. Flow velocity at the confluence of the superior vena cava to the right atrium up to 3 m/s, the calculated pulmonary artery systolic pressure was 70 mm Hg. Volume overload and dilatation of the right ventricle, and the compressed left ventricle were seen on transthoracic echocardiography (Fig. 1). Computed tomography performed shortly thereafter, clarified the anatomy of the echocardiographic findings (Fig. 2, Fig.3).

The presence of a wide patent ductus arteriosus and restrictive patent foramen ovale, contributed to hemodynamic instability. Despite that the child was on a constant prostaglandin E1 infusion, and cardiotonic support, his condition was progressively worsening: arterial hypoxemia developed (SaO₂-60%), also with combined arterial hypotension with systolic blood pressure of 30 mm Hg, and the pCO₂ increased up to 75 mm Hg.

An emergency operation was performed on the first day of life after the informed consent was obtained. We performed a median sternotomy, and moderately hypothermic cardiopulmonary bypass was established with ascending aorta and bicaval cannulations. After CPB initiating and cooling the patient’s body to 28°C, aorta was clamped and antegrade blood cardioplegia was used for cardiac arrest. Afterwards the aortic cannula was advanced to the brachiocephalic trunk and fixed with a tourniquet, the left common carotid and subclavian arteries were clamped,
and brain perfusion was started. CPB flow reduced to 30% of initial flow of 200 ml/min/kg. Patent ductus arteriosus was ligated and divided. The descending aorta was clamped distal to the narrowing. A resection of the narrowed portion of the aorta was performed – extended anastomosis was formed under the arch of the aorta. Thereafter the aortic cannula was repositioned to the ascending aorta, and the clamps were removed from the carotid and subclavian arteries. The duration of selective brain perfusion was 23 min. Full flow CPB was continued and the patient was warmed up. Correction of the TAPVC was performed using the “sutureless” technique [1]: an anastomosis was formed between the left atrium appendage and the pulmonary veins collector with a continuous stitch. After aortic clamp was removed, cardiac activity was restored without any arrhythmias. Aortic clamping time was 49 min, CPB time (with selective brain perfusion) was 83 min. Modified ultrafiltration was used after cardiopulmonary bypass. The chest was closed primarily. Ultrasound episcan of the heart realized in the operating room showed the blood flow velocity through the pulmonary veins’ collector was less than 1 m/s, and the peak gradient did not exceed 10 mm Hg at the level of the proximal aortic anastomosis, right ventricular pressure reduced to 30 mm Hg. The postoperative period was uneventful, the time spent in the intensive care unit was three days. At the time of discharge, the condition of the patient after 12 days was regarded as satisfactory. 2 months after surgery, the patient has good heart function, heart failure is absent, there is no re-coarctation and he has a good outflow through the pulmonary veins.

## Conclusion

Acute heart failure, shock and acidosis often develop around 8–10 days of life in children with complex coarctation [2]. In an international population-based study evaluated with 422 cases, TAPVC and aortic coarctation co-existence was observed only in six (1.4%) cases. Only several cases were reported of repair combination of different types of anomalous pulmonary venous drainage and coarctation of the aorta in newborns and children [3-6]. De Leval et al. reported in 1973 a case of successful repair of a supracardiac non-obstructive total anomalous pulmonary venous drainage and preductal coarctation of the aorta in a 12-days old newborn [7]. We have presented the case of successful repair of combination of obstructive TAPVC and coarctation of the aorta of one-day-old newborn.

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## Conflict of Interest Statement

We have no conflict of interest to declare. Authors have nothing to disclose with regard to commercial support.

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Figure 1. TTE before surgery. Enlarged right ventricle (RV) compresses the left ventricle (LV). Right ventricular end diastolic size (RVEDS) is 11 mm, left ventricular end diastolic size (LVEDS) is 9.5 mm, indexed left ventricular diastolic volume (iLVEDV) is 5 ml/m², left ventricular ejection fraction (EF LV) is 60%
Figure 2. CT scan before surgery. Preductal coarctation of the aorta is marked by the arrow.
Figure 3. CT scan before surgery. Totally anomalous pulmonary veins connection into the vertical vein. Collector stenosis marked by the arrow.