Cor Triatriatum Dextrum: A Case Report

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Abstract

This paper describes a diagnosis of and surgical treatment of Cor Triatriatum Dextrum in a term baby who, six hours postpartum, was determined to be cyanotic. The diagnosis was reached using two-dimensional echocardiography and angiocardiography. This case is the second diagnosis of cor triatriatum dextrum made during the first day of life, and the first successful surgery of this type performed on a newborn.

Introduction

Cor triatriatum dextrum is a rare defect that consists of abnormal septation of the right atrium. It is caused by persistence of the right valve of the sinus venosus, which normally regresses by week 12 of fetal development, leaving as its only full-term remnants the crista terminalis (superiorly) and the eustachian thebesian valves (inferiorly).1,3,14

Materials and Methods

A 3.5 kilogram, six-hour-old, cyanotic infant was brought to the Operating Room from the Cardiac Catheterization Laboratory on an urgent basis. A diagnosis of Persistent Right Valve of the Sinus Venosus had been made in the cath lab. The chest was entered through a median sternotomy, and the aorta and right atrial appendage were cannulated with a 2.5 mm cannula and a 4 mm cannula respectively. The patent ductus arteriosus was ligated and the patient was placed on cardiopulmonary bypass.4 The patient’s body surface area was .23M2; the pre-pump arterial blood gas on 100% oxygen was 7.41 pH; the pCO2 was 33; the pO2 was 46; base excess was +0.7; and the O2 saturation was 92 percent.

The extracorporeal circuit pump was primed with 250 cc of crystalloid and 500 cc of whole blood, to which sodium bicarbonate (20 mEq.) and heparin (2,000 units) were added. One-quarter-inch I.D. venous and arterial lines were used.

The patient was surface-cooled to a temperature of 32°C rectal, then heparinized with 300 units per kilogram. Upon the start of bypass, the patient was cooled to an average temperature of 16°C. During the cooling and rewarming phases, pulsatile flow was used. We use a maximum Cardiac Index of 3.2 L/min/M2 for our infants. Some difficulty with the systemic venous return occurred, and only half-flow was obtained. After two minutes of cardiopulmonary bypass, the venous cannula was changed to a 5 mm cannula. This did not improve the flow, and the venous return improved only after repositioning the cannula inside the right atrium. After eight minutes of cooling, the patient’s circulation was arrested. Acid base balance was managed by using the “Alpha Stat” theory of blood gas management.

Upon opening the right atrium, a web-like membrane was found arising laterally at the crista terminalis and

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on the floor of the right atrium, leaving the coronary sinus to the right and the inferior vena cava to the left. The superior vena cava was also to the left of the membrane, and at this level the structure had many fenestrations, which explains the difficulties with the venous return. This anatomy corresponds to the type III classification of cor triatriatum dextrum as described by Doucette and Knoblich.3

The membrane was resected. The tricuspid valve was normal. The atrial septal defect (ASD) was visible, the pulmonary venous return was normal, and the defect was repaired. The right atrium was closed, cardiopulmonary bypass restarted, the patient rewarmed, and the CPB terminated uneventfully.

Early in the post-operative course the patient developed nodal bradycardia which responded to small doses of Isuprel. Twelve hours post-op she was again in sinus rhythm. The patient was discharged, in good condition, on the eighth post-operative day.

Discussion

All case reports before 1970 of “Cor Triatriatum Dextrum” or “Persistent Right Valve of the Sinus Venosus” were identified at autopsy as noted by Thomka, et al.12 Another case was reported by Gerlis, et al.4

The diagnosis of this defect remains difficult because of its rarity. Since 1970, six cases have been reported; two diagnosed by echocardiography6,7,10,13 One case did not go to surgery.13 In three other cases,8,9,12 cor triatriatum dextrum was discovered during surgery, the preoperative diagnosis having been tricuspid stenosis in two patients, and “pendulous” myxoma of the right ventricle in the other.

Summary

The patient described in this paper is the first one to survive the operation at one day of age. From the perfusion standpoint, this case was interesting as an example of complications that can occur during cardiopulmonary bypass as a result of anatomical considerations which are unknown until the heart is opened, and therefore cannot be anticipated by the perfusionist.

References