Negative results - Congenital

Pentalogy of Cantrell associated with hypoplastic left heart syndrome and herniation of the ventricular mass into the abdominal cavity

James D. St. Louis
Division of Cardiac Surgery, Medical College of Georgia, 1120 15th St., BA-4300, Augusta, Georgia 30912-4040, USA

Received 20 September 2005; received in revised form 10 February 2006; accepted 13 February 2006

Abstract

Objective: A term female with the diagnosis pentalogy of Cantrell associated with hypoplastic left heart syndrome was born without complication. A significant portion of the sternal body was hypoplastic. The right ventricle was found to be herniated into the abdominal cavity. Method: The patient underwent an uncomplicated repair of an omphalocele. The infant then underwent an uncomplicated modified Norwood procedure. The right ventricular mass was reduced into the thoracic cavity prior to the Norwood. Results: Because of inability to close the anterior chest wall, a skin allograft was employed to cover the heart. The child died of fungal sepsis at 3 months of life. Conclusions: Repair of these complex constellation of defects, although successful technically, should take into account long-term morbidity that precludes a positive outcome.

Keywords: Allograft; Congenital heart disease; Hypoplastic left heart syndrome; Norwood; Sternum

1. Introduction

Pentalogy of Cantrell was initially described by Cantrell in 1958 [1]. The syndrome consists of (1) a defect in the lower sternum, (2) a supraumbilical abdominal wall defect, (3) a deficiency of the anterior portion of the diaphragm, (4) a deficiency in the diaphragmatic portion of the pericardium, and (5) a congenital heart defect. The cardiac defect most often involves a ventricular septal defect, an atrial septal defect, double outlet right ventricle, tetralogy of fallot, or a left ventricular diverticulum [2]. Most cases are sporadic, although there has been an association with an X-linked inherited mutation [3]. Survival rate for patients with pentalogy of Cantrell is low, with the outcome dependent on the complexity of the cardiac defect.

2. Materials and methods

The patient for presentation is a 2.8-kg black female. She had a known in utero diagnosis of hypoplastic left heart syndrome (HLHS) and an uncharacterized midline abdominal wall defect. At birth, the patient was stabilized on a prostaglandin infusion and electively intubated. Physical examination revealed severe hypoplasia of the distal sternal body and xiphoid. There was an omphalocele with reducible abdominal contents. ECHO evaluation confirmed the diagnosis of HLHS.

The patient was initially taken to the operating room on the 5th day of life for repair of the abdominal wall defect. During exploration, the ventricular mass was found herniated through a deficiency in the diaphragm (Fig. 1). The omphalocele was repaired primarily, making no attempt at reducing the heart. The infant made an uneventful recovery.

The infant was taken to the operating room on the 11th day of life and underwent a Mee modification of the Norwood procedure. During initial exploration, a large portion of the right ventricle was found to be herniated through a moderate-sized defect in the diaphragm. The...
heart was easily reduced without hemodynamic compromise. The infant was weaned from cardiopulmonary bypass without complication. Because of vertical orientation of the right ventricle, the sternum was stented open and covered with a silastic membrane.

3. Results

The post-operative course was uncomplicated. The patient was taken to the operating room four times over the ensuing ten days for wash out and attempted closure of the sternum. Each attempt to reduce the heart into the pericardial cavity was met with severe hemodynamic instability, requiring immediate reopening. Several unsuccessful attempts were made to reconstruct the hypoplastic anterior chest wall. Because of the inability to reduce the heart into the pericardial cavity, a skin allograft (Alloderm™) was used to close the defect. Over the next two and half months, the patient was periodically taken to the operating room for reduction of the skin patch (Fig. 2). Approximately 1 cm was taken from the skin allograft edge at each procedure. Unfortunately, the patient expired from a fungal sepsis likely related to prolonged intra-venous access.

In conclusion, this report describes the rare combination of severe hypoplasia of the sternum with herniation of a single ventricle into the abdomen. Reduction of the heart and correction of the cardiac defect was without complication. The lack of a sternal body, hypoplasia of the pericardial cavity, and new orientation of the ventricular mass never allowed adequate closure of the sternal defect.

References