Ann Thorac Surg 1997;64:836-838

Case Report

Anomalous Origin of Left Coronary From Right Pulmonary Artery in Hypoplastic Left Heart Syndrome

George E. Sarris, MD, Jonathan J. Drummond-Webb, MD, Makram R. Ebeid, MD, Larry A. Latson, MD, Roger B. B. Mee, FRACS

Center for Pediatric and Congenital Heart Surgery, The Cleveland Clinic Foundation, Cleveland, Ohio

Accepted for publication April 8, 1997.

Abstract

An infant with hypoplastic left heart syndrome presented for surgical repair at 9 months of age, the ductus having remained open in the presence of a restrictive atrial septal defect. In addition, an anomalous left coronary artery originating from the right pulmonary artery was found. After preliminary blade/balloon atrial septostomy, a successful modified Norwood procedure with concomitant reimplantation of the anomalous coronary artery was performed.

Top
Footnotes
Abstract
Introduction
Comment
References

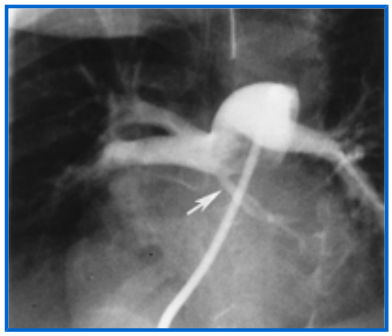
Introduction

Without treatment, hypoplastic left heart syndrome (HLHS) is usually rapidly fatal either as a result of ductal closure and systemic hypoperfusion or due to overwhelming heart failure. Rarely, however, survival may be prolonged if the ductus remains open and a restrictive atrial septal defect sufficiently limits pulmonary blood flow. In infants with anomalous origin of the left coronary artery from the pulmonary artery, the anomalous coronary artery usually originates from the main pulmonary artery. Origin from the right pulmonary artery has been reported in only 3 patients [1–3]. We report our experience with a rare case of combined HLHS and anomalous left coronary artery originating from the right pulmonary artery

Top
Footnotes
Abstract
Introduction
Comment
References

presenting for surgical management at 9 months of age.

The patient, born with HLHS variant, had the diagnosis made prenatally. Although the parents initially chose "compassionate care," the child survived. However, growth and development were poor, there was marked tachypnea, and the family decided to pursue surgical therapy at 8 months of age. Repeat evaluation revealed hypoplastic left ventricle and mitral valve with a 6.5-mm ascending aorta but hypoplastic transverse arch, coarctation, large patent ductus arteriosus, bilateral superior venae cavae, and a perimembranous ventricular septal defect. Patency of the ductus had permitted survival, but the lungs were markedly congested. A restrictive atrial septal defect was present [4], left atrial pressure was 30 mm Hg, and pulmonary artery pressure was systemic. In addition, the left coronary artery originated anomalously from the proximal right pulmonary artery (Fig 1). Pulmonary vascular resistance was high at 15 Wood U/m^2. To assess potential lability of pulmonary resistance, we performed limited blade/balloon atrial septostomy. Pulmonary blood flow increased (pulmonary-to-systemic flow ratio rose from 1:1 to 2.5:1) and pulmonary vascular resistance decreased to 8 Wood U/m^2. This suggested that pulmonary resistance could drop further after complete left atrial decompression, increasing the chance of operability. Therefore, we decided to proceed with a modified Norwood procedure [5] and concomitant reimplantation of the anomalous coronary artery.



View larger version (144K):
[in this window]
[in a new window]

*Fig 1. . Anteroposterior projection of pulmonary angiogram demonstrating the origin of the left coronary artery (*arrow*) from the proximal right pulmonary artery.*

Under hypothermic cardiopulmonary bypass, the distal branch pulmonary arteries were snared to preserve perfusion to the anomalous coronary. Under cardioplegic arrest, the ductus was ligated, the main pulmonary artery was transected just proximal to the bifurcation, and the anomalous coronary artery was excised from the right pulmonary artery with a generous cuff and translocated to the back wall of the proximal aorta. The cross-clamp was removed, all coronary arteries filled well, the heart started beating slowly while cooling continued, and the distal main and proximal right pulmonary arteries were repaired with autologous pericardium. At a nasopharyngeal temperature of 18°C, circulatory arrest was instituted and a modified Norwood procedure was performed as previously described [5].

Postoperative recovery was uneventful. Four months later, room air arterial saturation was 75% and follow-up catheterization demonstrated good ventricular function with no outflow obstruction and normal filling of the reimplanted coronary artery. On the basis of persistently moderately elevated pulmonary vascular resistance (3.3 Wood U/m^2) responsive to oxygen, the patient was maintained on home oxygen to be reevaluated for possible bidirectional Glenn shunt in a few months.

Comment

Top

Footnotes
Abstract
Introduction
Comment
References

Anomalous origin of the left coronary artery from the right pulmonary artery is a very rare anomaly [1–3], the repair of which by direct aortic implantation has only recently been reported [1]. Furthermore, we found only one autopsy report of combined HLHS with an anomalous coronary artery [6]. Survival beyond a few weeks of life in patients with HLHS receiving compassionate care alone is also quite rare, and in our case this became possible by a persistently patent ductus preserving systemic perfusion, in combination with a restrictive atrial septal defect limiting pulmonary blood flow. The resultant pulmonary hypertension could have precluded surgical palliation due to development of pulmonary vascular obstructive disease [4], but it did preserve perfusion of the anomalous coronary artery and ventricular function. Limited blade/balloon atrial septostomy demonstrated lability of the pulmonary vascular resistance and suggested operability. Combined repair involving coronary translocation as well as modified Norwood procedure [5] was successful, although future conversion to cavopulmonary shunt (and, ultimately, a Fontan circulation) still remains uncertain.

Footnotes

Top
Footnotes
Abstract
Introduction
Comment
References

Address reprint requests to Dr Sarris, Department of Pediatric and Congenital Heart Surgery, The Cleveland Clinic Foundation, 9500 Euclid Ave, Cleveland, OH 44195.

References

Top
Footnotes
Abstract
Introduction
Comment
References

1. Tanaka SA, Takanashi Y, Nagatsu M, Ohta J, Hoshino S, Imai Y. Origin of the left coronary artery from the right pulmonary artery. Ann Thorac Surg 1996;61:986–8.[Abstract/Free Full Text]
2. Brodman RF, Issenberg HJ, Glickstein JS, Frame R. Use of a free radial artery graft for correction of Bland-White-Garland syndrome. Ann Thorac Surg 1996;62:1525–6.
3. Doty DB, Chandramouli B, Schieken RE, Lauer RM, Ehrenhaft JL. Anomalous origin of the left coronary artery from the right pulmonary artery. Surgical repair in a 10 month old child. J Thorac Cardiovasc Surg 1976;71:787–91.[Abstract]
4. Canter CE, Moorhead S, Huddleston CB, Spray TL. Restrictive atrial septal communication as a determinant of outcome of cardiac transplantation for hypoplastic left heart syndrome. Circulation 1993;88(Pt 2):456–60.
5. Fraser CD, Mee RBB. Modified Norwood procedure for hypoplastic left heart syndrome. Ann Thorac Surg 1995;60:S546–9.
6. Ito T, Nuno M, Isiukawa J, et al. Hypoplastic left heart syndrome with a single coronary artery originating from the pulmonary artery. Acta Pediatr Jpn 1995;37:60–3.

This article has been cited by other articles:

E. Villa, G. Brancaccio, A. Carotti, P. Francalanci, and R. M. Di Donato
Circumflex Coronary Artery From Right Pulmonary Artery in Hypoplastic Left Heart Syndrome
Ann. Thorac. Surg., November 1, 2005; 80(5): 1919 - 1920.
[Abstract] [Full Text] [PDF]

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY HOME

M. Nosal', I. C. Omeje, and R. Poruban
Hypoplastic left heart syndrome with anomalous origin of left coronary artery from the right pulmonary artery: successful surgical treatment in a neonate
Eur. J. Cardiothorac. Surg., September 1, 2005; 28(3): 497 - 498.
[Abstract] [Full Text] [PDF]

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY HOME

G. Oppido, C. P. Napoleone, S. Martano, and G. Gargiulo
Hypoplastic left heart syndrome in situs inversus totalis
Eur. J. Cardiothorac. Surg., November 1, 2004; 26(5): 1052 - 1054.
[Abstract] [Full Text] [PDF]

THE ANNALS OF THORACIC SURGERY HOME

J. J. DeRose Jr, R. Corda, M. R. Dische, J. Eleazar, and R. S. Mosca
Isolated left ventricular ischemia after the Norwood procedure
Ann. Thorac. Surg., February 1, 2002; 73(2): 657 - 659.
[Abstract] [Full Text] [PDF]

THE ANNALS OF THORACIC SURGERY HOME

E. Malec, T. Mroczek, J. Pajak, A. Zajac, J. Kolcz, and K. Januszewska
Hypoplastic left heart syndrome with an anomalous origin of the left coronary artery
Ann. Thorac. Surg., December 1, 2001; 72(6): 2129 - 2130.
[Abstract] [Full Text] [PDF]

Articles by Sarris, G. E.
Articles by Mee, R. B. B.
Search for Related Content

PubMed

PubMed Citation
Articles by Sarris, G. E.
Articles by Mee, R. B. B.