AORTIC ATRESIA WITH VSD/Thiene et al.

References


Anatomy of Aortic Atresia

Cases Presenting with a Ventricular Septal Defect

GAETANO THIENE, M.D., VINCENZO GALLUCCI, M.D., FERGUS J. MACARTNEY, M.D., STEFANO DEL TORSO, M.D., PIERO A. PELLEGRINO, M.D., AND ROBERT H. ANDERSON, M.D.

SUMMARY The anatomy of 58 specimens of aortic outflow tract atresia was studied. All cases had situs solitus and levocardia, 37 had atroventricular (AV) concordance, two had common inlet to a right ventricle and 19 had mitral atresia. The great arteries were normally interrelated in all cases. Fifty-one cases had an intact ventricular septum, while seven presented with a ventricular septal defect (VSD). Of the seven with VSD, in two it was associated with a common AV orifice draining exclusively into the right ventricle in the presence of a rudimentary left ventricular chamber. In one case a small VSD accompanied combined mitral and aortic atresia. In the other four cases the left ventricles and mitral valves were fairly normal in size; the VSD was subpulmonary in three cases, due to infundibuloventricular malalignment, and perimembranous in one. These last four cases are of particular interest since they could be amenable to surgical correction. Possible approaches to surgical treatment and morphologic features pertinent to them are described and discussed.

MANY INVESTIGATORS have studied aortic atresia and have revealed a high degree of uniformity in morphology.\(^1\)\(^-\)\(^4\) Recently, variations of morphology have been described; aortic atresia was found in hearts with ventricular septal defects (VSD) and normally developed left ventricles.\(^5\)\(^-\)\(^12\) This reported variation in morphology, some of it of potential surgical significance, prompted us to review the anatomy of hearts with aortic atresia, with special reference to cases with VSD.

From the Departments of Pathology, Pediatrics and Cardiovascular Surgery, University of Padova, Medical School, Padova, Italy, and from the Department of Pediatrics, Cardiothoracic Institute, Brompton Hospital and the Thoracic Unit, The Hospital for Sick Children, London, England.

Address for reprints: Dr. Gaetano Thiene, Department of Pathology, Via Gabelli 61, 35100 Padova, Italy.


Materials

Fifty-eight hearts with aortic atresia were obtained from the Department of Pathology at the University of Padova, Italy; the Thoracic Unit, The Hospital for Sick Children, London, England; and the Department of Pediatrics, Cardiothoracic Institute, Brompton Hospital, London, England.

All hearts were from persons with vissemi-atrial situs solitus and levocardia. In 37 hearts, we observed atrioventricular (AV) concordance, with the left atrium and the left ventricle connecting through a hypoplastic mitral valve. In 19 hearts, there was absence of the left AV connection (mitral atresia). In the remaining two hearts, both atria drained through a common AV orifice to a right ventricle,\(^13\) a rudimentary chamber of left ventricular type being identified in both. In all hearts the hypoplastic ascending aorta was posterior and to the right of the pulmonary trunk.
Fifty-one hearts had an intact ventricular septum and seven had a VSD. Among the latter, one with an apical VSD had combined mitral-aortic atresia and hypoplastic left ventricle. In another two hearts, the VSD was part of a dominant right type of common AV orifice with rudimentary left ventricular chamber. Finally, four had VSD and normally developed left hearts.
Anatomy of Hearts with VSD and
Normally Developed Left Ventricle and Mitral Valves

Four hearts had normally developed left ventricles and mitral valves. Quantitative analysis, calculated for the age in the manner described by Lev et al., disclosed that the left ventricle was dilated with normal thickness in two specimens. Another heart exhibited pressure hypertrophy with normal volume of the left ventricular cavity. Finally, one specimen showed a concentric left ventricular hypertrophy with reduced cavity due to the association with a restrictive VSD (fig. 1). The mitral orifice was normal in three hearts and slightly reduced in one.

The pulmonary trunk arose entirely from the right ventricle in three hearts (figs. 2 and 3) and overrode the ventricular septum in the other (fig. 1). The VSD was subpulmonary in three specimens, with the posterior rim formed by a muscular structure which separated the pulmonary and AV valves (figs. 1 and 2), probably representing the posteriorly-displaced infundibular septum. In another heart, the infundibular septum was normal in size and position, with the large defect situated at the junction of the membranous and muscular components of the ventricular septum (fig. 3). An imperforate aortic valve membrane was present in this last specimen.

One heart had bicuspid pulmonary valve (fig. 2) and one a dysplastic pulmonary valve (fig. 1). Discrete coarctation was observed in three hearts, one preductal and two juxtaductal.

One specimen had an anomalous origin of the right subclavian artery distal to the left subclavian artery. Two other hearts showed bilateral ductus arteriosus. The right patent ductus joined the right pulmonary artery to the right subclavian artery in one heart and to the brachiocephalic artery in the other (fig. 1). In this last heart the brachiocephalic artery was not connected with the ascending aorta.

Discussion

The morphologic findings of aortic atresia are similar to those of pulmonary atresia. As previously stated by Roberts et al. and Lev, the hearts can function with a VSD or with an intact ventricular septum. The finding of some hearts with adequately developed left ventricles suggests the possibility for future surgical intervention.

Aortic atresia associated with an adequately developed left ventricle and mitral valve has recently been recognized as a discrete anatomic-clinical entity. It was first described by Lev and subsequently by Rosenquist et al. in a case presenting with an aortopulmonary fenestration and interrupted aortic arch. Only later was its significance realized and its clinical and surgical importance emphasized. This condition, which is relatively favorable for surgery, accounts for only a small proportion of all cases of aortic atresia. Roberts et al. reviewing 73 necropsy cases, found three hearts with normal left ventricles and mitral valves (4%). The same incidence was
FIGURE 3. Aortic atresia with ventricular septal defect (VSD) and normally developed left ventricle and mitral valve. Top) Right ventricular view: note the perimembranous VSD with a normally sized and positioned infundibular septum (IS) and with tricuspid valve apparatus inserted in the roof of the defect (arrow). Bottom) The left ventricle (LV) is dilated and the mitral valve (MV) is normal in size. The atresia is at the valvular level (arrow) and the ventricular septal defect is the only outlet.
reported by Freedom et al. (six cases among 148, 4%). In our investigation, a slightly greater percentage has been found (four cases among 58, 7%). In this condition, the left ventricle and mitral valve were adequately developed, the left heart could be considered for reconstruction in each case. The presence of the VSD accounts for the normal development of the left chambers. If the presence of a normal left ventricle is established, either by angiocardiography or echocardiography, production of persistent patency of the ductus (obtainable with formalin infiltration) and banding of the pulmonary arteries will be the required surgical procedures for early palliation. Complete correction by separating the pulmonary and systemic circulations and establishing direct continuity between the heart and the aorta may be planned for a later stage.

In cases with normal mitral valves, two main surgical procedures may be used for total repair. Bernhard's technique has been used in case of severe stenosis of the aortic outflow tract, placing a conduit between the left ventricular apex and the descending aorta. When used for aortic atresia with VSD, this procedure requires closure of the septal defect together with ligation of the ductus. The brachiocephalic and coronary arteries would then be supplied retrogradely from the conduit flow. An alternative to this procedure would be to use the pulmonary trunk as the left ventricular-aortic conduit (fig. 4). This could be achieved by: 1) an intraventricular conduit to direct left ventricular blood through the VSD into the pulmonary artery; 2) a longitudinal incision of the main pulmonary artery and ductus arteriosus, with patch closure of the orifices of the right and left pulmonary arteries, and enlargement of the ductus with a longitudinal patch; and 3) establishing continuity between the right ventricle and the distal left and right pulmonary arteries with a T- or Y-shaped valved conduit.

While Bernhard's intervention is simpler and is not dependent on systemic circulation via the ductus, the presence of aortic coarctation, seen in three of our four cases, might interfere with retrograde blood supply to the brachiocephalic and coronary arteries. This could also present a problem with the second procedure, when the coarctation is preductal. Freedom et al. suggested anastomosing the pulmonary trunk into the side of the ascending aorta. In any case, coarctation of the aorta requires precise clinical definition and careful surgical attention. Moreover, difficulties arise in incorporating the pulmonary trunk 1) if it originates predominantly from the right ventricle, 2) if the VSD is small, or 3) if the tricuspid apparatus inserts directly into the superior rim of the VSD.

Acknowledgment

The authors thank Dr. Carlo Marcelletti for the suggestions on the surgical approach.

References

thirty autopsy cases in infants with surgical considerations. Am J Cardiol 21: 166, 1968
Anatomy of aortic atresia. Cases presenting with a ventricular septal defect.
G Thiene, V Gallucci, F J Macartney, S Del Torso, P A Pellegrino and R H Anderson

Circulation. 1979;59:173-178
doi: 10.1161/01.CIR.59.1.173

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1979 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/59/1/173

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/