appropriate, and perhaps even life-saving, to start a
PGE{subscript}_2 infusion before transfer of an infant to a center
for definitive diagnostic evaluation by echocardiography and cardiac catheterization. However,
systemic output also may be improved in infants with
hypoplastic left heart syndrome; this has been our ex-
perience. Since clinical differentiation between
hypoplastic left heart syndrome and other forms of
left ventricular outflow obstruction is often difficult, it
may be preferable to improve systemic blood flow
temporarily in an occasional infant with the
hypoplastic left heart syndrome than delay this in in-
fants with surgically correctable lesions such as coar-
tation or interruption of the aorta.

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Anatomy of Aortic Atresia
Cases Presenting with a Ventricular Septal Defect

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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 in-
tact 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 the remaining 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 sub-
pulmonary 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. 1-14 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.

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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

All hearts were from persons with viscero-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 rudimen-
tary 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.
FIGURE 1. Aortic atresia with ventricular septal defect and normally developed left ventricle and mitral valve. A) External view: the brachiocephalic artery (BA) arises from the right pulmonary artery, via a right ductus arteriosus, without exhibiting confluence with the ascending aorta (AA). PT = pulmonary trunk. B) Right ventricular view: the pulmonary trunk overrides the ventricular septal defect and shows dysplastic pulmonary valve. IS = infundibular septum. C) The left ventricle (LV) exhibits concentric hypertrophy, the left ventricular cavity is slightly reduced and the mitral valve (MV) is normal. The ventricular septal defect (VSD) is small, its posterior wall being formed by the posteriorly deviated infundibular septum.

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 angiocardiology 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.

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