Complete Common Atrioventricular Canal in Infancy — Surgical Repair and Postoperative Hemodynamics

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DOLORES VITULLO, M.D., JOHN LAMBERTI, M.D., RENE A. ARCILLA, M.D.,
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SUMMARY Fourteen infants with complete common atrioventricular canal (CCAVC) underwent open heart surgery under deep hypothermia and circulatory arrest. There were three operative deaths and two late deaths. Postoperative studies performed in seven of the nine survivors revealed nearly normal hemodynamics. There were no residual shunts, and excellent mitral valve function was observed in six patients. In one patient, residual mitral regurgitation was noted. The pulmonary artery pressures and pulmonary vascular resistances were normal except in one who had severe pulmonary vascular obstructive disease before surgery. The mean left ventricular end-diastolic volume changed from 175 ± 24% (SEM) before surgery to 106 ± 7% after surgery (P < 0.01). The corresponding right ventricular end-diastolic volume changed from 166 ± 16% to 102 ± 19% (P < 0.025). Left ventricular ejection fraction was mildly decreased before and after surgery (0.63 ± 0.02).

Surgical repair of CCAVC is possible during the first year of life, with likely normalization of cardiac size and function. Unsatisfactory results related to pulmonary vascular obstruction may be anticipated if repair is delayed much beyond the first year.

THERE HAVE BEEN SEVERAL preliminary reports on the surgical repair of complete common atrioventricular canal (CCAVC) during infancy. However, excluding our series, the follow-up has usually been limited to clinical assessment of results, with the exception of two patients who underwent postoperative cardiac catheterization. This report describes our experience since January 1974 in the surgical repair of CCAVC in 14 infants. The postsurgical hemodynamics of seven of the nine survivors are also described.

Materials and Methods

Fourteen infants (five females, nine males) with CCAVC and no associated major intracardiac anomalies underwent open heart surgery. All were in cardiac failure, and two required mechanical ventilatory support for several days before surgery. One other infant had two episodes of cardiopulmonary arrest. Thirteen of the 14 had Down’s syndrome. At the time of surgery, their ages ranged from 3-15 months (mean 8 months), and their weight ranged from 3.4-6.2 kg (mean 4.7 kg). Only two patients were more than 1 year of age (13 and 15 months; see table 1). Six patients had patent ductus arteriosus. One patient without Down’s syndrome had situs inversus, dextrocardia and azygous continuation of the inferior vena cava.

Preoperative Cardiac Catheterization

Preoperative hemodynamic data were obtained an average of two months before surgery (table 2). One patient had diagnostic studies at another institution. All patients had pulmonary hypertension with the pulmonary/aortic systolic pressure ratio ranging from 0.45–1 (mean 0.90). In the patient with dextrocardia and azygous continuity of the inferior vena cava (case 4), the pulmonary artery pressure was not obtained. However, the right ventricular systolic pressure was 90% of systemic pressure, and there was no pulmonary stenosis confirmed in the postoperative studies. Assumed oxygen consumption (162 ml/min/m2) was used to calculate blood flows by the Fick method. The calculated pulmonary and aortic blood flows showed considerable variation, as did the respective vascular resistances. The pulmonary/systemic flow ratios ranged from 0.8–4.9 (mean 2.3). The pulmonary/systemic resistance ratios ranged from 0.12–1.20 (mean 0.50).

None of the 14 patients had hypoplasia of either ventricle, and in all instances the balanced form of CCAVC, according to the criteria of Bharati and Lev, was observed. Cardiac chamber volumes were calculated from the biplane cineangiograms and compared with the predicted normal based on the patient’s age, height, weight and heart rate. In 10 patients where the right ventricular size could be determined, the end-diastolic volume ranged from 118%–275% of normal (mean 196%). In all 13 patients where the left ventricular size could be calculated, the end-diastolic volume ranged from 95%–282% of normal (mean 158%). The left ventricular ejection fraction, obtainable only from 11 patients, ranged from 0.51–0.67 (mean 0.61).
Surgical Technique

Open heart surgery was carried out using either anesthesia, surface or perfusion cooling for profound hypothermia, and circulatory arrest for periods ranging from 40–68 minutes (mean 54 min) at reciral temperatures of 15°C–20°C (mean 18°C).

The surgical anatomy in three patients satisfied the Rastelli criteria for a type A defect, and in eight patients for a type C defect (see discussion). In the remaining three patients, the intracardiac anatomy did not fit into any of the three types described by Rastelli et al.14 The surgical repair was based on the method described by Rastelli and colleagues.15, 16 Both the atrial and ventricular portions of the septal defect were closed with a single dacron patch using superficial interrupted mattress sutures along the right side of the muscular septum to secure the patch. In most patients, both the anterior and posterior bridging leaflets had to be split in order to expose the large interventricular communication for placement of the patch. A greater portion of the bridging leaflet tissue was assigned to the mitral valve and was used to reconstruct this valve. The medial part of the mitral valve was secured to the dacron patch at a level 6–8 mm superior to the crest of the ventricular septum, thus elevating it well above the left ventricular outflow. One patient required right ventriculotomy to secure the anterior aspect of the patch while in the remainder, the repair was accomplished using only a right atrial approach. In 13 patients, simultaneous pulmonary artery and aortic pressures were obtained intraoperatively after completion of the intracardiac repair, warming and establishment of sinus or AV junctional rhythm. Blood samples for oxygen saturation were also obtained from the superior vena cava, pulmonary artery and systemic artery.

Results

Overall Mortality

Three infants died in the early postoperative period, for an operative mortality of 21% (table 1). Two of these deaths were attributed to dehiscence of the suture lines used to reconstruct the mitral valve. After an uneventful recovery, case 1 (age 5 months) suddenly deteriorated three days after surgery, and emergency cardiac catheterization confirmed severe mitral regurgitation. At reoperation, the suture lines of the mitral valve "cleft" repair had torn out, as did the approximation of the anterior leaflet to the dacron patch. The patient died one week after secondary repair of the dehiscence. Another child (case 11) also showed sudden clinical deterioration on the third postoperative day, accompanied by a drop in cardiac output monitored by thermodilution. At autopsy, one of three mitral sutures was found to have dehisced. The third early death (case 6) exhibited persistently low cardiac output after surgery, and died on the second postoperative day. Autopsy was refused.

There were two late deaths. In one infant, death occurred six weeks after surgery and was attributable to pneumonia and sepsis. At necropsy, the operative repair appeared intact. The other patient, who was clinically well and in sinus rhythm at the time of discharge and at the first follow-up clinic visit, was dead on arrival at another hospital three months after surgery. There was no autopsy, and the cause of death is unknown. Including the two late deaths, the overall mortality was 36%.

Follow-Up

The follow-up in one baby recently operated on has thus far been limited to six weeks postsurgery. Eight late survivors have been followed for up to 36 months, and in all but one good cardiopulmonary status has been maintained. In the only exception (case 2), mitral regurgitation has persisted; and although the heart size has regressed, he continues to be on digoxin therapy. The other eight patients have normal auscultatory findings with either no murmur or only a faint, short, nonspecific ejection systolic murmur and a well-split second heart sound with normal or slightly accentuated pulmonary component.

High degree atrioventricular block occurred late in two cases: In one patient complete AV block appeared at four months (case 3), and in another patient second-degree AV block (Mobitz type 2) appeared at 14 months postoperatively (case 2). Both had permanent pacemaker implantation. Another patient has intermittent AV dissociation that has not progressed further and has been well-tolerated for two years (case 4). Six patients remain in sinus rhythm with normal PR intervals.

Post-Operative Cardiac Catheterization

Follow-up cardiac catheterization has been carried out four to 14 months following surgery in seven of the nine survivors. Figure 1 shows the serial changes in the pulmonary arterial-to-systemic peak systolic pressure ratios obtained preoperatively, immediately after the surgical repair, and at the second catheterization study. The pulmonary artery pressures obtained intraoperatively in 13 patients all declined. These decreased further or remained essentially unchanged at subsequent catheterization. Moderate pulmonary hypertension was encountered in the one case who had advanced peripheral pulmonary vascular obstruction preoperatively (case 8). His pulmonary vascular resistance (465 dyne-sec-cm⁻⁵/m²) remained elevated

<table>
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<th>Table 1. Patient Summary: Age at Surgery and Outcome</th>
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although significantly lower than that before surgery (710 dyne-sec-cm⁻²/m²). In the other six patients, the pulmonary vascular resistances were normal (mean 150 dyne-sec-cm⁻²/m²) (fig. 2).

The changes in the configuration of the left ventricle following the surgical repair as seen angiographically are shown in figure 3. Before surgery, the typical gooseneck deformity was observed in all cases, accompanied by left-to-right shunting at the ventricular level and varying degrees of AV valve regurgitation into the left and right atria. After surgery, the gooseneck deformity was significantly less since the deficient portion of the atrioventricular septum was occupied by the patch and the mitral valve shifted to a more cephalad level. Biplane left ventricular angiography confirmed functional integrity of the mitral valve in six of the seven patients. Mitral regurgitation was demonstrated postoperatively in only one patient, discussed below.

The pre- and post-operative ventricular end-

![Figure 1](http://circ.ahajournals.org/)

**Figure 1.** Serial changes in the pulmonary artery-to-systemic artery peak systolic pressure ratios obtained preoperatively, immediately after surgical repair, and at the postoperative catheterization study.
diastolic volumes, expressed in percent of predicted normal, appear in figure 4. Significant reduction in left ventricular and right ventricular size was observed following surgery. One patient (case 2) showed slight increase in left ventricular size. This child had residual mitral regurgitation, with a regurgitant fraction of 20% calculated by our angiographic method. This was the same case in whom second degree AV block appeared 14 months postoperatively. His clinical status deteriorated at that time, and the left ventricular end-diastolic volume as well as left atrial maximal volume were 140% and 130% of the predicted normal, respectively. Restoration of a more physiologic ventricular rate following pacemaker implantation resulted in clinical improvement. His left ventricular and left atrial dimensions, obtained in subsequent echocardiograms, have decreased, although they remain slightly greater than normal. Although the mean right ventricular end-diastolic volume of the seven postsurgical patients was normal (102% ± 19 SEM), the right ventricular size in one patient (case 7) increased slightly, from 143% to 168% of normal.

Table 3 is a summary of the hemodynamic findings before and after surgery for the seven patients restudied. Group analysis by t test revealed significant reduction of the pulmonary artery pressure,
pulmonary flow, pulmonary/systemic resistance ratio, and right and left ventricular end-diastolic volumes. The end-diastolic pressures of both ventricles remained slightly elevated or at high normal levels in some of the patients postoperatively. There was no evidence for mitral stenosis after surgery. The mean pulmonary capillary wedge pressure obtained from four patients was 11 mm Hg ± 0.5 (SEM), and these did not differ significantly from the simultaneously recorded left ventricular end-diastolic pressures. In
Fig. 4. Pre- and postoperative ventricular end-diastolic volumes, expressed in percent of predicted normal. Normal range is represented by the shaded area.

Discussion

Based on the combined experiences of several centers in recent years,5, 6, 9 the operative mortality from the surgical repair of CAVC during infancy is 20–25%. With increasing experience in operative techniques and in patient selection, a decrease in operative risk can be expected. Thus, only one early death occurred in the 1976 series of Kirklin and associates consisting of 10 patients under two years of age.6

The cardiac anatomy is such that surgical repair is difficult. The mitral components of the common AV valve continue into the tricuspid components as anterior bridging and posterior bridging leaflets with varying patterns of chordal attachments. The leaflets may be thin and delicate or thickened and, occasionally, myxomatous-like. Rarely, there is considerable deficiency in leaflet tissue, and this will compromise the repair. Occasionally, the valve leaflets tend to be preferentially distributed into one ventricle which is small. Bharati and Lev11 have referred to such cases showing hypoplasia of one ventricle as dominant right or dominant left types. We also believe that such rare cases are not correctable not only because of the smallness of one ventricle, but also because of the unusual chordal attachments of the leaflets.

The atrial and ventricular components of the septal defect in CAVC vary in size, although both are usually significantly large. If the interatrial communication is small, as occasionally occurs, the exposure to the mitral valve is difficult unless the atrial septum is incised. A high incidence of associated cardiac lesions is observed, especially when splenic abnormalities are also present. These include coarctation of the aorta, patent ductus arteriosus, tetralogy, double-outlet right ventricle, single ventricle, positional abnormalities of the heart and others. Splenic abnormalities are likely to be observed in the patients without Down's syndrome.11 A child with CAVC and Down's syndrome is thus less likely to have other complicating cardiac lesions than the patient without this chromosomal disorder.

Rastelli and colleagues14 have classified CAVC into three types, based chiefly upon the configuration and chordal attachments of the anterior bridging leaflet: type A, with the anterior leaflet divided at the midportion and the chordae attached to both sides of the ventricular septum; type B, divided anterior leaflet with chordae unattached to the septum but attached medially to an anomalous papillary muscle in the right ventricle; type C, undivided anterior leaflet with chordae unattached to the septum but attached to the usual papillary muscles in both ventricles. Despite its simplicity and usefulness for clinicopathologic studies, this classification is not applicable for all hearts. Thus,
three of our cases could not be so categorized. In addition, the demarcation between categories, especially between the A and C types, is not always sharp.11

The amount of AV valve tissue available for mitral valve reconstruction, especially that of the anterior bridging leaflet, is critically important to the surgical repair. An incompetent mitral valve is likely to result if there is not enough tissue from which to fashion an adequate, functional mitral anterior leaflet.

Mitral regurgitation was observed in only one of our long-term survivors, and none showed clinical or hemodynamic evidence for mitral stenosis. However, acute mitral regurgitation secondary to dehiscence of the mitral valve sutures appeared to have caused two surgical deaths. This complication has not been previously emphasized as a cause for surgical failure. Additional considerations relevant to the surgical management are the presence of other intracardiac anomalies, the status of the myocardium, the extent of pulmonary vascular obstruction and the vulnerability of the conduction system.

High degree AV block as a late complication of CCAVC repair has not been reported by others. This occurred twice in our series, early in our experience (second and third cases) and has not been observed subsequently. Interestingly, these were the only two patients who had not converted to sinus rhythm within the first week after surgery although both exhibited sinus rhythm with prolonged PR interval upon their discharge. The anatomy of the His bundle in this condition, specifically, its intimate relationship to the posteroinferior rim of the septal defect in the ventricular septum, is well established.17,18 Trauma to this critical site during patch placement may cause transient or permanent AV block.

The natural history of CCAVC is characterized by early death from congestive heart failure.19,20 Without some type of early surgical intervention, only about one-third of the patients survive the first year. In recommending primary open heart repair of CCAVC during the first year of life, Mair and McGoon5 cited two reasons for this approach: 1) ineffective palliation and high mortality from pulmonary artery banding, and 2) high risk of secondary peripheral pulmonary vascular obstruction occurring in unoperated patients after 1 year of age. Although some children with CCAVC do improve following pulmonary artery banding alone, it is not possible to identify with certainty which patients are likely to respond favorably to this palliative procedure and which ones will not.

Marked clinical improvement has been reported after early primary repair.5,6 However, objective assessment of the hemodynamics after surgery has been lacking. Espulgas et al.7 found normal ventricular end-diastolic pressures in a group of children who had CCAVC repair at an older age (including two who had pulmonary artery banding before age 1 year). Although the left ventricular end-diastolic volumes estimated from single plane angiograms remained elevated, stroke volumes increased proportionately as normal ejection fractions were maintained. Residual shunting or mild-to-moderate mitral regurgitation was observed in five of the eight patients, and these probably accounted for the persistently increased left ventricular volumes.

Our experience suggests that nearly normal hemodynamics can be achieved by early primary repair of CCAVC in a majority of patients who would otherwise not survive the first year. Intracardiac shunting diminishes, and restoration of normal mitral valve function is possible. Left ventricular size becomes normal unless residual volume overload is
present, as was noted in one of our patients who had persistent mitral regurgitation. The pulmonary hypertension and high pulmonary vascular resistance are generally reversible. The exception in our series was the oldest in the group (case 8), referred to us after 1 year of age and operated on at age 15 months, despite severe peripheral pulmonary vascular obstruction.

Cordell et al.21 have demonstrated the return of left ventricular size and function to normal following closure of large ventricular septal defects during the first two years of life. The mean left ventricular ejection fraction for their 13 patients was normal before and after surgery. However, the mean left ventricular ejection fraction for our patients was below normal before and after CAVC repair. It would thus appear that the left ventricular volume overload in our patients with CAVC resulted in impaired pump performance at a very early age. In view of the changes in left ventricular loading following surgery (increased afterload and decreased preload), it is possible that myocardial contractility has somewhat improved postoperatively although ejection fraction remains mildly reduced. We do not have sufficient information to predict whether ventricular function will improve further with time. Serial assessment of resting and exercise hemodynamics is indicated in these and in similar groups of patients.

Acknowledgments

The authors wish to acknowledge with thanks the continued support of Mrs. Arma Wyler and of the Home for Destitute Crippled Children.

References

15. Rastelli GC, Ongley PA, Kirklin JW, McGoon DC: Surgical repair of the complete form of persistent common atrioven-

The Morphologic Spectrum of Double Outlet Left Ventricle and Its Surgical Significance

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HUGH A. McALLISTER, JR., M.D., AND JOHN W. KIRKLIN, M.D.

SUMMARY Double outlet left ventricle (DOLV) is defined in this paper as that condition in which both great arterial vessels emerge completely (complete form) or mostly (incomplete form) from the morphologic left ventricle. This is a study of eight cases diagnosed during surgery and 37 cases diagnosed at autopsy. DOLV was classified as follows: 1) with the aorta to the right, or to the right and posterior to the pulmonary trunk: a) with ventricular septal defect (VSD) confluent with the aorta (10 cases), b) with VSD confluent with the pulmonary trunk (three cases), and c) with VSD confluent with both vessels (two cases). 2) with aorta anterior to, anterior and to the right, or anterior and to the left of the pulmonary trunk: a) with the VSD related to the aorta (10 cases), b) with the VSD related to the pulmonary trunk (one case), and c) with the VSD related to both vessels (two cases). 3) with tricuspid valve abnormalities: a) with tricuspid atresia and stenosis with normally related vessels (four cases), or aorta anterior (10 cases), b) with straddling or displaced tricuspid valve (two cases), or c) with Ebstein's anomaly (one case). Five surgical and four autopsied cases were of the complete form. From the surgical standpoint, it is important in DOLV to determine the interrelationship and origin of the great arteries, their relationship to the VSD, and the location of the VSD in the ventricular septum. The presence of tricuspid valve abnormalities is emphasized.

We undertook combined pathologic and surgical study of DOLV to further elucidate the anatomic facts which might be surgically important.

Materials and Methods

This study consisted of a gross pathologic examination of 24 hearts from the Congenital Heart Disease Research and Training Center and 13 hearts from the Armed Forces Institute of Pathology. The study also included a review of the morphology of the eight cases (seven of whom survived operation and are doing well) operated on from 1967–1978 at the University of Alabama Medical Center, Birmingham, Alabama. Only hearts with atrioventricular (AV) concordant connections were included in the study.

The atria and viscera were in situ solitus in each case. In the pathologic material all hearts were in levocardia. Two of the surgical cases had dextroversion.

The presence or absence of tricuspid valve anomalies was determined for each heart. The abnormalities included tricuspid stenosis, tricuspid atresia, displaced tricuspid orifice and Ebstein's anomaly. In displaced tricuspid orifice, both the mitral and tricuspid orifices enter the left ventricle completely.
W Culpepper, J Kolff, C Y Lin, D Vitullo, J Lamberti, R A Arcilla and R Replogle

Complete common atrioventricular canal in infancy--surgical repair and postoperative hemodynamics.

Circulation. 1978;58:550-558
doi: 10.1161/01.CIR.58.3.550

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1978 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

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