Pediatric Cardiac Surgery Annual

COMPLETE ATRIOVENTRICULAR CANAL

Overview: History, Anatomy, Timing, and Results of Complete Atrioventricular Canal

Carl L. Backer, Robert D. Stewart, and Constantine Mavroudis

The past 50 years have been marked by major advances in the care of children with complete atrioventricular canal defects. There have been important contributions from surgeons, cardiologists, and pathologists to provide us with our current understanding of the anatomy of atrioventricular canal defects and excellent surgical outcomes. In this monograph we will discuss the surgical contributions of Lillehei, Kirklin, McGoon, Maloney, Trusler, Wilcox, and Nunn. The improvements in outcomes achieved by these surgeons were made possible by the pathology and anatomy clarifications provided by Lev, Rastelli, and Anderson.
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KEYWORDS AV septal defect, endocardial cushion, AV valve

The mean age at the time of surgical repair for children with complete atrioventricular (AV) canal has dropped from 6 years of age in 1959 to currently 3 months of age. Surgical mortality has incrementally dropped from over 50% to less than 5%. The use of transesophageal echocardiography has improved our understanding of the left AV valve repair and attention to the "zone of apposition." The repair of an AV canal defect still has an Aristotle score of 9 (range, 1.5 to 15), which means congenital heart surgeons consider this one of the more complex lesions that undergoes repair. The purpose of this article is to provide an overview of the history, anatomy, timing of surgery, and surgical results for infants with complete AV canal.

History

The first successful repair of an AV canal defect was reported by C. Walton Lillehei in 1955.[1] He used cross-circulation and direct suture of the atrial rim of the septal defect to the crest of the ventricular septum. In the late 1950s and early 1960s Lillehei, Kirklin, McGoon, and Cooley converted to the use of cardiopulmonary bypass and used two separate patches of Ivalon or Teflon "sponge" to close the atrial and ventricular components of the defect separately.[2-4] Suture closure of the "mitral valve cleft" was a standard part of the repair. Maloney reported the use of a single patch (Dacron) technique in 1962.[5] In 1976, George Trusler reported a two-patch technique with prosthetic material for the ventricular septal defect and a pericardial patch for the atrial component.[6] Dwight McGoon reported successful surgical repair of AV canal during the first year of life in 1977.[7] He recognized the importance of "taking from the tricuspid valve" to leave sufficient tissue from which to create an adequate left AV valve. Carpentier believed that the left AV valve functions best when repaired as a 3-leaflet valve.[8] This opened the debate on how to manage the mitral valve cleft, now called the "zone of apposition."[9] Ben Wilcox and Graham Nunn independently reported a "simplified" or "modified" single patch technique for repair of AV septal defect in 1997 and 1999, respectively.[10,11]

Anatomy

The AV canal defects, also referred to as AV septal defects and endocardial cushion defects, encompass a wide spec-

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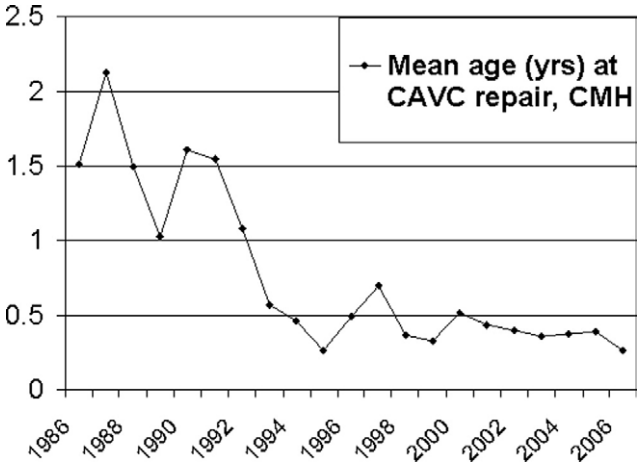

Figure 1 Chart showing the mean age of complete AV canal repair at Children's Memorial Hospital from 1986 to 2006.

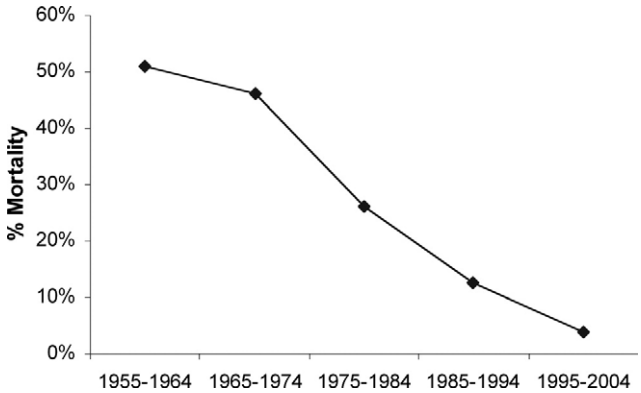

Figure 3 Mortality rate shown from a meta-analysis of surgical results from 1955 to 2004.

trum of anatomic findings. In addition to the complete AV canal defect, there are the partial atrioventricular canal (AVC) (ostium primum atrial septal defect) and intermediate (transitional) AVC.[12] The anatomy of the complete AV canal defect is extremely important in determining the type of repair and outcomes. In 1958, Maurice Lev described the position of the AV node and the bundle of His.[13] This helped surgeons avoid third degree AV block. The first classification of the anatomy of complete AV canal was by Giancarlo Rastelli.[14] Rastelli divided complete AVC into three major groups, now known as Rastelli A, B, and C. Professor Robert Anderson emphasized that the cardinal feature that distinguishes the "AV Canal" is its common AV junction.[15] Associated with this common junction is the "unwedged aorta" and discrepancy between the inlet and outlet dimensions of the ventricular mass.[16]

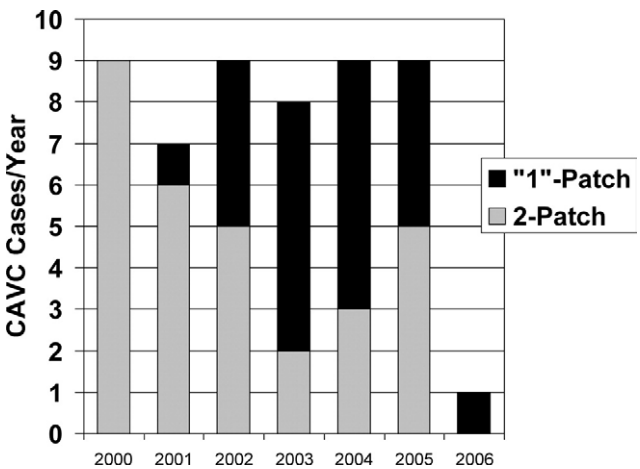

Figure 2 Chart depicting the transition from the two-patch technique to the modified single-patch technique for complete AV canal repair at Children's Memorial Hospital from 2000 to 2006.

Timing of Operative Interventions

Over the past 50 years the age at which complete AV canal is electively repaired has dropped. The Mayo Clinic (Rochester, MN) series published in 1959 had 15 patients with a median age of 6 years, (range, 10 months to 16 years).[3] The change in mean age at the time of complete AV canal repair at Children's Memorial Hospital (Chicago, IL) from 1986 through 2006 is shown in Fig 1.[17] In the past 2 years at Children's Memorial Hospital the mean age at the time of repair of patients with a complete AV canal has been 4.5 months. In Nunn's series, median age was 3.4 months.[11] The results of AV canal repair have actually improved as the age of the patients at the time of surgery has dropped. The initial drop in mortality was mostly secondary to avoidance of complications of pulmonary hypertension. More recent improvements are related to operating before the development of pneumonia or other complications related to congestive heart failure and AV valve insufficiency. The ability to perform these repairs in early infancy has been facilitated by improvements in anesthesia, cardiopulmonary bypass, and postoperative ICU management. The use of the "simplified" or "modified" single-patch technique has also facilitated repair in early infancy. At Children's Memorial Hospital we actually appear to be transitioning to favoring the "modified" single-patch technique, as evidenced in Fig 2. Most centers now perform AV canal repair with only a preoperative transthoracic echocardiogram on which to base the diagnosis. Routine cardiac catheterization is no longer used.[18] The ideal age for complete AV canal repair appears to be 3 to 6 months of age.

Table 1 Results: Classic Single-Patch Technique

Institution	Year	No. of Patients	Operative Death	Mitral Insufficiency	Third degree AV Block
Boston Children's[20]	1992	301	3%*	9%	3%
UCLA[21]	1990	105	7%†	6%	2%

*Since 1987.
†Since 1986.

Table 2 Results: Two-Patch Technique

Institution	Year	No. of Patients	Operative Death	Mitral Insufficiency	Third degree AV Block
Children's Memorial	2005	173	10 (6%)	14 (8%)	7 (4%)
Munich[22]	1998	251	18 (7%)	31 (12%)	10 (4%)
Berlin[23]	1996	120	12 (10%)	7 (6%)	4 (3%)
Indianapolis[24]	1995	203*	16 (8%)	8 (4%)	5 (2%)

*Switched to two-patch technique in 1987.

Table 3 Results: Simplified Single Patch

Institution	Year	No. of Patients	Operative Death	Mitral Insufficiency	Third degree AV Block
Australia[11]	1999	47	2 (4%)	1 (2%)	0
Children's Memorial	2006	22	1 (4%)	1 (4%)	0
North Carolina[10]	1997	12	1 (8%)	0 (0%)	0

THE DIRECT-VISION INTRACARDIAC CORRECTION OF CONGENITAL ANOMALIES BY CONTROLLED CROSS CIRCULATION

RESULTS IN THIRTY-TWO PATIENTS WITH VENTRICULAR SEPTAL DEFECTS, TETRALOGY OF FALOT, AND ATRIOVENTRICULARIS COMMUNIS DEFECTS

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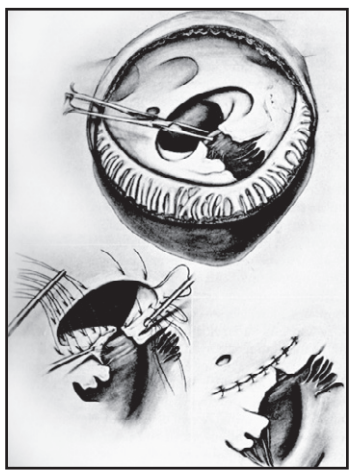
Surgery 1955;38:11-29[1]

C. Walton Lillehei, MD



Operative technique for repair of atrioventricularis communis, complete form

(Artist's drawing dated June 1954)

Figure 4 First successful repair of an AV canal defect – cross circulation. (C. Walton Lillehei, 1955.)

Surgical Results

The results of complete AV canal repair have improved dramatically over the past 50 years. Lillehei reported 9 deaths out of 20 patients (45% mortality) in his original series.[2] The first Mayo Clinic series reported a mortality rate of 73% (11 deaths out of 15 patients) in 1959.[3] Denton Cooley reported 16 deaths out of 24 patients (67%) in 1960.[4] Current results with the three most commonly used different techniques are shown in Tables 1, 2, and 3.

In the current era, mortality is now less than 5% in nearly all series. The mortality rate in a meta-analysis calculated from a large number of surgical results is shown in Fig 3. Surgical efforts initially aimed for survival and the avoidance of heart block and ventricular level shunts. As results improved, the surgical focus shifted to the left-sided AV valve. Left AV valve insufficiency has been one of the major causes of reintervention in these patients. The incidence of reoperation for AV valve insufficiency currently ranges from 5% to 10%. Most surgeons perform routine closure of the left AV valve "zone of apposition."[9] The incidence of third degree AV block has also dropped, and in most series this now ranges between 1% and 4%.

Two techniques have been described during the atrial repair to avoid AV block. Some surgeons keep the coronary sinus on the right atrial side and carry the atrial suture line adjacent to the mitral valve. Other surgeons suture the atrial patch in such a manner that the coronary

Maurice Lev, M.D.

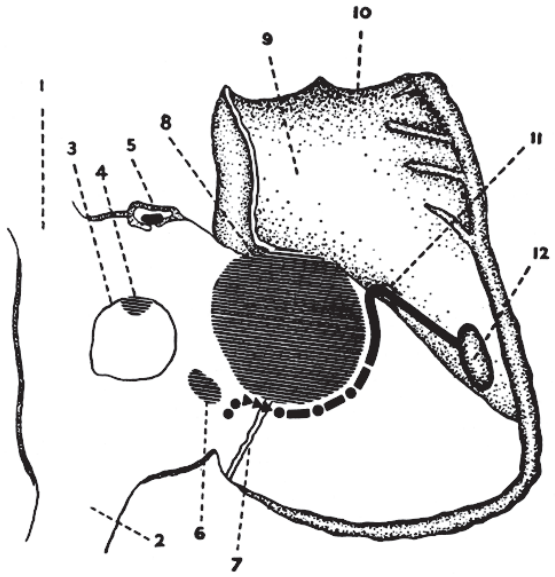


Fig. 12.—Diagrammatic sketch of the course of the AV node, bundle, and right bundle branch in the anomaly common atrioventricular orifice. Right atrial and ventricular view. Dots indicate AV node; triangles, penetrating portion of the AV bundle; dots and dashes, branching portion of the atrioventricular bundle; solid line, right bundle branch; 1, superior vena cava; 2, inferior vena cava; 3, limbus; 4, patent foramen ovale; 5, cut edge of atrial appendage; 6, entry of coronary sinus; 7, base of tricuspid valve; 8, combined patency of atrial and ventricular septa; 9, conus; 10, base of pulmonary valve; 11, muscle of Lancisi; 12, cut edge of moderator band.

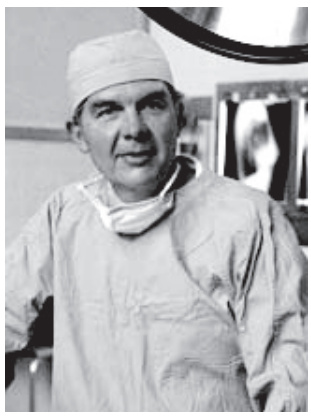
Figure 5 Lev's description of the AV node and the bundle of His.

sinus drains to the left atrial side. The incidence of AV block comparing these two techniques does not appear to be different. Intraoperative transesophageal echocardiography is now routine in the evaluation of residual atrial or ventricular level shunting or valve insufficiency at the time of initial repair. In the recent Children's Memorial series, eight patients out of 120 (6.6%) underwent immediate intraoperative re-exploration with cross-clamp and cardioplegia for the above noted residual lesions.

Conclusion

The monumental contributions of surgical pioneers such as Lillehei, Kirklin, Cooley, McGoon, Maloney, Rastelli, Wilcox, and Nunn, along with Maurice Lev's studies on the conduction system have led to the excellent results with repair of AV canal that we have today (Figs 4-10). Improvements in patient management strategies now occur in small but important increments. An example is the "simplified" or "modified" single patch recently added as a "new" operation to the "classic" single- and double-patch procedures used over the past 40 years. These operations remain challenging, as evidenced by the complete AV canal Aristotle score of 9 (range, 1.5 to 15.0).[19] Congenital heart surgeons consider complete AV canal repair more difficult than ventricular septal defect repair (score, 6.0), but less difficult than truncus arteriosus repair (score, 11.0). Surgeons will continue to evaluate the outcomes of different surgical techniques for different anatomic defects, and use that information to revise and optimize surgical results.

John W. Kirklin, MD



Dwight C. McGoon, MD

THE SURGICAL TREATENT OF
ENDOCARDIAL CUSHION DEFECTS

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Surgery **1959;46:185-196**

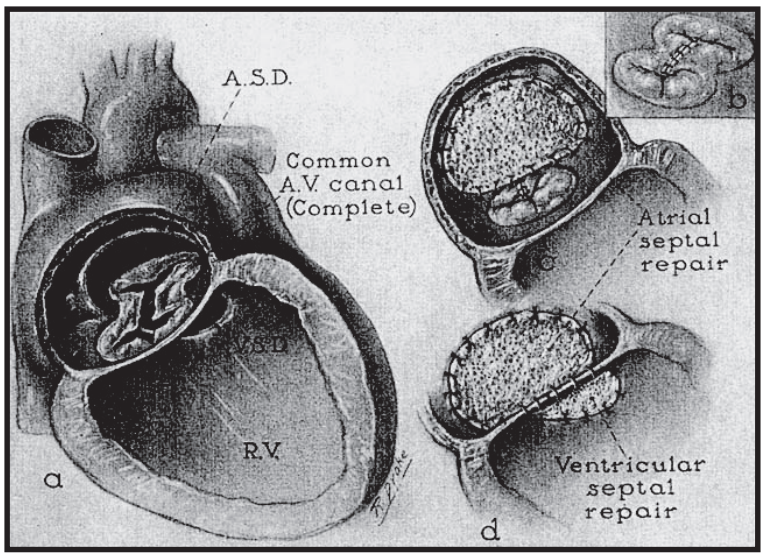

Figure 6 Repair of AV canal with cardiopulmonary bypass and two Teflon sponges.

The Surgical Treatment of Common Atrioventricular Canal

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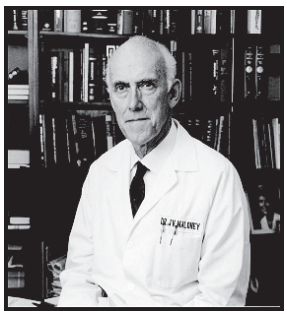
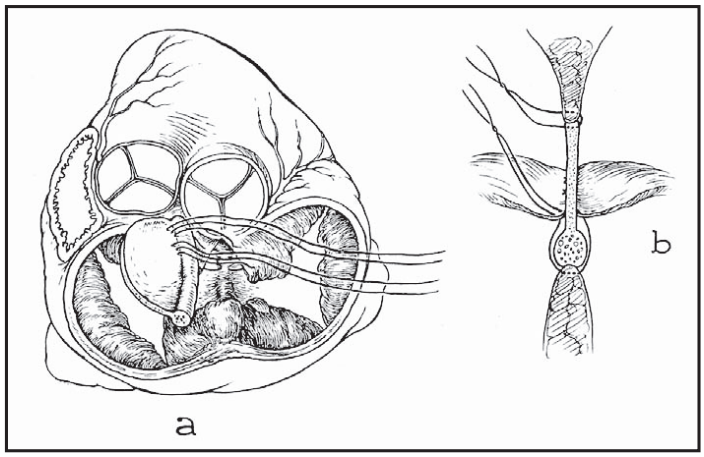
J Thorac Cardiovasc Surg **1962;43:84-96**



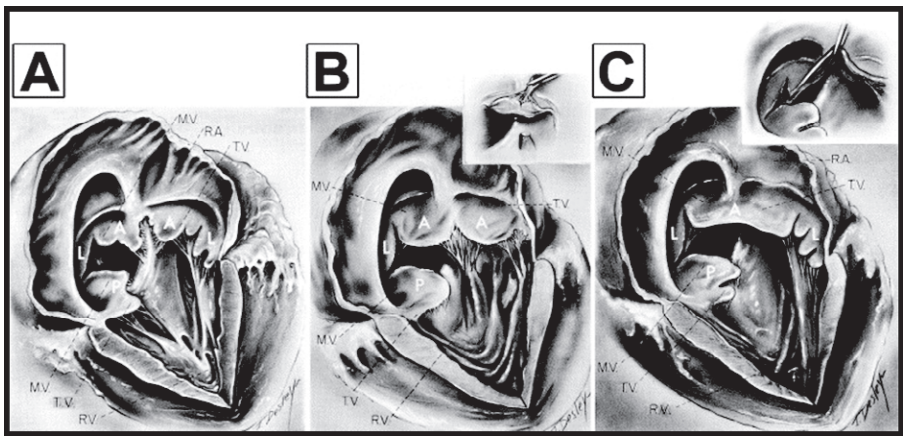
James V. Maloney, Jr. MD



Repair by splitting the anterior common atrioventricular valve leaflet to avoid excessive valve deformity deformity by downward displacement. (a) View from atria showing configuration of prosthesis and course of sutures. (b) Final relation of split valve leaflets to the prosthesis.

Figure 7 Single patch technique. James Maloney, 1962.

Giancarlo Rastelli, MD



Rastelli classification of complete common atrioventricular canal. (A) Type A: Anterior (A) common atrioventricular leaflet is divided into two portions, one mitral valve (MV) and one tricuspid valve (TV), attached medially to interventricular septum with long, nonfused chordae tendineae. In posterior common AV leaflet, MV and TV portions are not separated. (L = lateral; P = posterior; RA = right atrium.) (B) Type B: Anterior common AV leaflet is divided but not attached to the septum. Mitral and tricuspid components are both attached medially to abnormal papillary muscle arising in right ventricle near septum. Free interventricular communication occurs under anterior common leaflet. (RV = right ventricle.) (C) Type C: Anterior common AV leaflet is not divided and is not attached to the septum so that free interventricular communication, extending to vicinity of aortic cusps, occurs underneath this leaflet. (From Rastelli GC, et al. Mayo Clin Proc 1967;42:200-9 [7])

Figure 8 First classification of the anatomy of complete AV canal by Giancarlo Rastelli.

Benson R. Wilcox, MD

Anatomically Sound, Simplified Approach to Repair of Complete Atrioventricular Septal Defect

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Background. There are few congenital anomalies of the heart that have benefited more from thorough Anatomic analysis than the complex anomaly known as atrioventricular septal defect in the setting of common atrioventricular junction. Recent advances in understanding the anatomy of this lesion have led to alternative methods of repairing these defects.

Methods. The medical records of 21 consecutive patients undergoing repair of complete atrioventricular septal defect have been reviewed. Nine of these Patients had a standard one- or two-patch repair, and 12 had direct closure ~~of the ventricular~~ element of the defect.

Results. Direct closure resulted in significantly shorter pump and cross-clamp times. Follow-up for an average of 34 months suggests that when direct closure can be performed, the results are comparable with those of the more standard technique.

Conclusions. Our initial success with this approach is encouraging; however, longer follow-up is required to establish whether it will be broadly applicable.

…direct closure of the ventricular element

(*Ann Thorac Surg* 1997;64:487-94)

Figure 9 Wilcox's description of the simplified single-patch technique.

SIMPLIFIED SINGLE PATCH TECHNIQUE FOR THE REPAIR OF ATRIOVENTRICULAR SEPTAL DEFECT

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Objective: Because of the complexity of traditional 1- and 2-patch techniques for the repair of complete atrioventricular septal defect, we modified our repair technique to avoid the use of any ventricular septal patch material. We report our prospective experience with this simplified 1-patch technique. ***Method:*** Forty-seven consecutive patients between May 1995 and August 1998 underwent repair with the use of this technique without modification. Repair was done in all patients by direct suturing of the common atrioventricular valve leaflets to the crest of the ventricular septum. No division of valve leaflets was necessary. A single pericardial patch was used to close the defect in the atrial septal component. Follow-up included electrocardiography and echocardiographic assessment of ventricular function, atrioventricular valve function, and adequacy of the left ventricular outflow tract. ***Results:*** There were 2 deaths (4%), only 1 cardiac related, in the series. There were 17 male patients and 30 female patients. Mean age at repair was 5.6 months (median, 3.4 months). Associated lesions were repaired in 19 patients (40%). Mean follow-up was 1.85 years (median, 1.9 years). There was no heart block. There were no significant residual ventricular septal defects detected and no left ventricular outflow tract obstruction seen on echocardiography in any patient to date. Mitral valve status after operation was assessed as no incompetence in 13 patients (28%), minimal in 19 patients (40%), mild in 12 patients (26%), and moderate in 3 patients (6%). ***Conclusion***: The repair of complete atrioventricular septal defect by direct suturing of the atrioventricular valve leaflets to the crest of the ventricular septum with a single-patch technique greatly simplified the repair and does not lead to left ventricular outflow tract obstruction nor interfere with valve function.

(*J Thorac Cardiovasc Surg* 1999;118:642-7)

Figure 10 Nunn's description of the simplified single-patch technique.

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