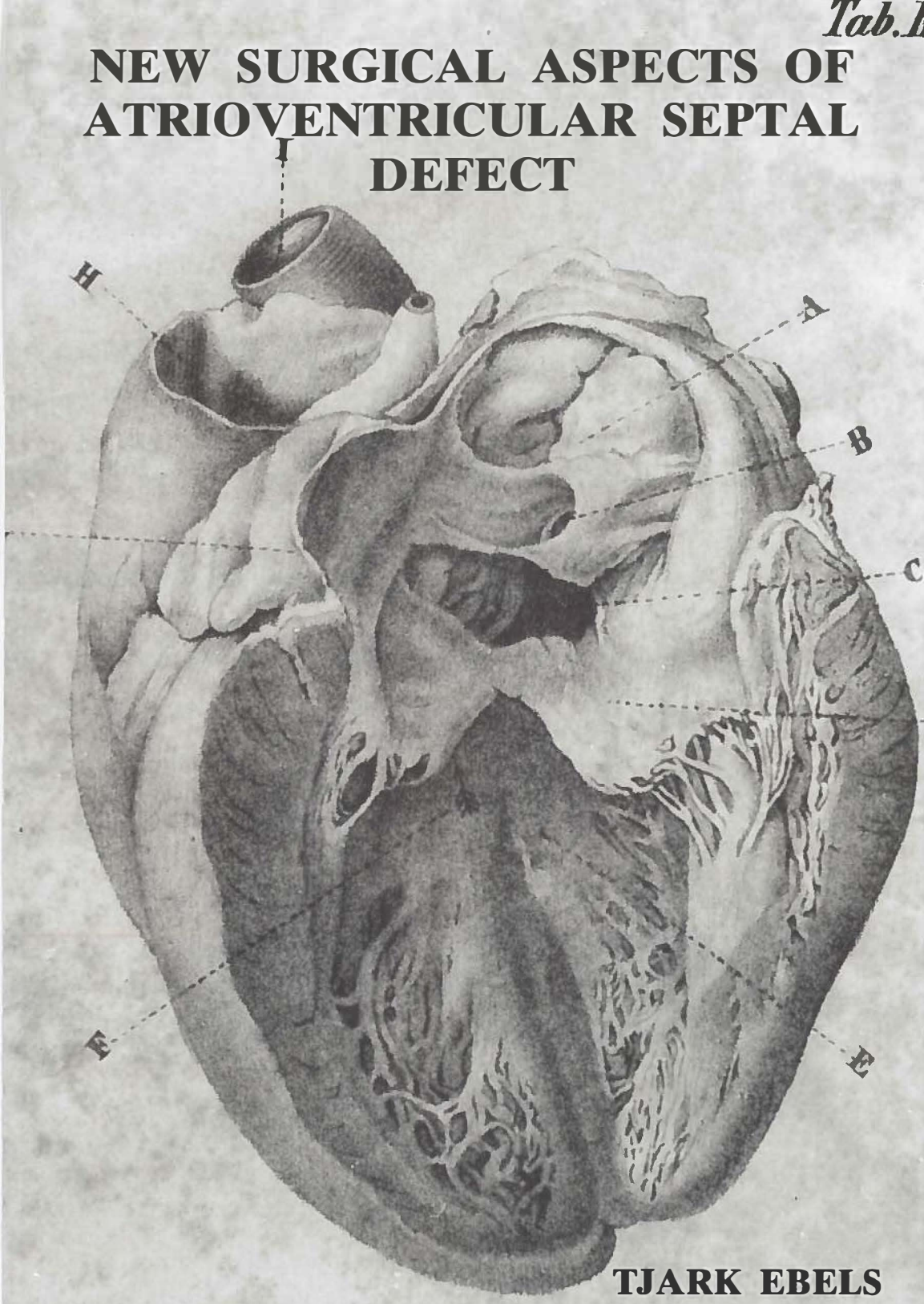
NEW SURGICAL ASPECTS OF ATRIOVENTRICULAR SEPTAL DEFECT



TJARK EBELS

NEW SURGICAL ASPECTS OF ATRIOVENTRICULAR SEPTAL DEFECT

STELLINGEN

1. Bij de vervanging van de linker atrioventriculaire klep bij een patient met een atrioventriculair septum defect, dient de achterwand van de linker ventrikel uitstroombaan gereceseerd te worden.

2. Het systolische geruis bij patienten met een atrioventriculair septum defect is vaak afkomstig van de linker ventrikel uitstroombaan en niet van een regurgiterende atrioventriculaire klep.

3. "Die Mitra des Bischofs kam in das Herz, wie der Teufel in den Weihbrunnkessel". Joseph Hyrtl, 1880.

4. Meting van de hoekgrootte van de drie bladen van de linker atrioventriculaire klep is van belang voor de operatieve benadering.

5. Het onderscheiden van een "intermediair type" atrioventriculair septum defect is zinloos en verwarrend.

6. Anatomische nomenclatuur dient vrij te zijn van embryologische smetten.

7. De demystificatie van de hartchirurgie krijgt financiële consequenties.

8. De ductus arteriosus, die Leonard Botal in 1565 beschreef als "vena arteriarum nutrix", was 1400 jaar eerder al door Claudius Galenus beschreven. De ductus verdient daarmee evenmin Botal's naam als Amerika die van Amerigo Vespucci.

9. Bij het bepalen van de behoefte aan open-hart-chirurgie is het zinloos ons te spiegelen aan de ons omringende in dezen nog minder ontwikkelde landen.

10. Over 10 jaar zullen minder dan de helft van het huidige aantal coronaire revascularisatie operaties worden uitgevoerd.

11. Afschaffing van de verplichte rechtsbijstand bij echtscheidingsprocedures is in strijd met de op overige gebieden van het recht bestaande tendens tot bescherming van de zwakkere partij.

12. Kunstgras is al dood.

13. Het voornaamste verschil tussen sportvliegen en open-hart-chirurgie is het slachtoffer

Stellingen behorende bij het proefschrift

New Surgical aspects of
Atrioventricular septal defect

Groningen, 3 mei 1989, Tjark Ebels

RIJKSUNIVERSITEIT GRONINGEN

NEW SURGICAL ASPECTS OF ATRIOVENTRICULAR SEPTAL DEFECT

PROEFSCHRIFT

ter verkrijging van het doctoraat

in de Geneeskunde

aan de Rijksuniversiteit Groningen

op gezag van de Rector Magnificus Dr. L.J. Engels

in het openbaar te verdedigen op

woensdag 3 mei 1989

des namiddags te 2.45 uur precies

door

TJARK EBELS

geboren te Haarlem

1989

DRUKKERIJ VAN DENDEREN B.V.

GRONINGEN

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Financial support by the following foundations for the publication of this thesis is gratefully acknowledged.

1. Foundation for the Development of Cardiopulmonary Surgery in Groningen.
2. Netherlands Heart Foundation.

Voor mijn Vader.

Bij het voltooien van dit proefschrift wil ik in de eerste plaats mijn ouders en mijn gezin danken voor de steun die ik van hen heb gekregen. Om dit te schrijven ben ik veelvuldig afwezig geweest, ik hoop dat de toekomst daar verandering in zal brengen. U, collegae van de afdeling Thoraxchirurgie, dank ik voor Uw welwillende houding als ik met "de wetenschap" in de weer was.
In het bijzonder geldt deze dank U, Hooggeleerde Homan van der Heide. Zonder Uw kennis, ervaring en werkkracht was ik geen thoraxchirurg geworden en was dit proefschrift niet ontstaan. Ik heb het als een voorrecht ervaren door U te worden opgeleid en tevens dat U mijn eerste promotor hebt willen zijn.
Professor Anderson, it has been an honour and a privilege to work with you in London as well as in Pittsburgh. Your tremendous knowledge on the morphology of congenital heart disease has been extremely stimulating for me scientifically as well as in practical surgery.
Hooggeleerde Eijgelaar, U dank ik bijzonder voor mijn opleiding in de kinder hartchirurgie. U hebt mij bovendien voortdurend in mijn wetenschappelijke activiteiten gestimuleerd.
Zeergeleerde Meijboom, U dank ik voor de prettige samenwerking die we tijdens een belangrijk deel van het tot stand komen van dit proefschrift hebben gehad. In samenspraak zijn we toch tot waarlijk originele ideeën gekomen.
Hooggeleerde Bossina, U dank ik voor de enthousiaste wijze van introductie in de kindercardiologie en voor de mogelijkheden die U mij geboden hebt.
Miek Schasfoort - van Leeuwen, jouw ijverige onbevooroordeeldheid is de basis geweest van twee hoofdstukken. Ik dank je hartelijk voor alle werk dat je verricht hebt en bovendien voor het onderricht in de echocardiografie dat ik spelenderwijs van je kreeg.
Hooggeleerde Kuipers, met U dank ik de hele afdeling kindercardiologie, die mij zo welwillend ten dienste was bij opzet en uitvoering van het onderzoek.
Siew Yen Ho, thank you very much for helping me to start off in the morphology of congenital heart disease and for the beautiful drawings that you made.
Hooggeleerde van der Wall, U dank ik voor de ideeën die culmineerden in mijn eerste artikel, de analogie met de HOCM blijft frappant.
Hooggeleerde Elema, U dank ik voor de fasciliteiten mij ter beschikking gesteld in het Pathologisch Anatomisch Laboratorium.
James R. Zuberbuhler, I thank you for giving me the opportunity to work in your department and for the hospitality that you bestowed upon me.
William A. Devine, Diane E. Debich, thank you for the ever friendly cooperation and all the unbiased measuring that you did in the Heart Museum.
L. Th. van der Weele, U dank ik voor het onderricht in de statistiek en het gebruik van de computer en voor alle hulp die ik bij problemen van U kreeg.
Cees Baarveld, David Brekhoff, Klaas Kooistra en André Winselaar, dank ik voor de assistentie op het Pathologisch Anatomisch Laboratorium.

Zeergeleerde Martis, met U dank ik Uw collegae Ackerman en Lo-A-Njoe, voor mijn opleiding in de algemene heelkunde, waar ik nog dagelijks plezier van heb.
Magda Munstra - Zuidema, Diny de Boer, Rina Timmer - Franssens en Marja de Bruin dank ik voor de secretariele hulp, zonder welke de manuscripten nooit zouden zijn verstuurd.

CONTENTS

CHAPTER I

INTRODUCTION

Anatomy

A heart with an atrioventricular septal defect exhibits not only a defect in a more or less two-dimensional septum, as is usually the case in a ventricular or atrial septal defect, but also possesses a cardiac architecture which differs completely from that of the normal heart[1]. Basically, the underlying anomaly is a defect of the inlet part of the ventricular septum. The atrioventricular valves would in normal circumstances, have derived, in part, from this deficient structure. Normal development, therefore, is impossible, which explains why these valves themselves develop in a characteristically abnormal way[2]. Because of the complexity of the lesion there has been, and still is, confusion and debate about its true nature and the relationships between the leaflets of the valves and the septal structures (Chapter VIII).
Judging from the profound differences with the normal heart, formation of this inlet portion of the ventricular septum is crucial for normal cardiac development. In addition to the defect at the anticipated site of the atrioventricular septum, the entire atrioventricular junction is a common one, resulting in the characteristically abnormal anatomy. The central fibrous body (that connects and separates the leaflets of the aortic, mitral and tricuspid valve in a normal heart) does not exist as such. Instead there is one atrioventricular junction, guarded by an essentially pentafoliate valve. The aortic valve is "unwedged", displaced antero-superiorly and is no longer in fibrous connexion with the crux of the heart. The subaortic left ventricular outflow tract, normally capacious, then becomes long and narrow. The atrioventricular valvar complex is displaced apically with the paradoxical result that the septal defect is located to various degrees within the atrium. The geometry of the ventricular mass is also profoundly different. The inlet and outlet dimensions more or less equal in the normal heart, are unequal with the outlet dimension being much larger than the inlet. All these features are the subjects of the various chapters of this thesis.

Incidence

An atrioventricular septal defect is a congenital cardiac anomaly that accounts for 2.4% - 7.4% of all cardiac anomalies[3-5]. The variability in reported incidences

may reflect differences in maternal age, because there is a direct relation between the mother's age and the incidence of Down's syndrome. Congenital heart disease is reported to occur in 23% - 56% of children with Down's syndrome. And about 25% - 50% of the children with Down's syndrome have a form of atrioventricular septal defect. Thus, having an extra copy of chromosome 21 apparently tends to disturb the normal process of septation[6].
As surgery for atrioventricular septal defect has become increasingly successful in the last two decades, recent studies have revealed that the offspring of parents, one of whom has undergone surgical correction of atrioventricular septal defect, have a much higher chance of having the same lesion. A recent review of the relevant literature by Nora and Nora revealed a 14% recurrence risk, when the affected parent was the mother[7]. In a survey of our own group of patients, we have found the same trend (unpublished observations). Therefore, we must expect the incidence of atrioventricular septal defect to rise, along with the maturation of our patients who have undergone successful operation.

History

An atrioventricular septal defect with separate left and right atrioventricular orifices used to be called an "ostium primum atrial septal defect". This convention underscores the interpretational problems of the anatomy, because later investigations have shown the primal atrial septum itself to be intact. The reason for the confusion is understandable, because the atrioventricular valves in this lesion are displaced apically, and are attached to the crest of the (deficient) ventricular septum. The inlet ventricular septal defect is, therefore, effectively located within the atrium. And because the defect is located "low" in the atrium septum, it appears to be the primary embryological septum that is deficient. The evolution of the anatomical perception from 1846 to the present is the subject of Chapter II.

Surgery

Surgery of atrioventricular septal defect started with "closed" (that is, without a heart-lung machine permitting intracardiac vision) operations, in which closure of the atrial septal defect was attempted. The results were usually less than satisfactory, because the closed techniques did not enable the surgeon to avoid the conduction tissues that closely border the defect, at a time when pacemakers were still science fiction, particularly for the pediatric age group[8]. But just as important, the closed techniques did not enable the surgeon to deal with the left

atrioventricular valve, which often regurgitates, and even more so after improper surgical management. The advent of open heart techniques gave the surgeon time to assess and, if necessary, repair the left atrioventricular valve. The history of surgical treatment is the subject of Chapter III.
The results of our own investigations into the long term follow-up of patients that had been operated for their "ostium primum defect" were primarily presented at a meeting of the Dutch Association of Thoracic Surgery on May 15, 1982[9] and later expanded (Chapter IV).

One of the conclusions of the abstract was:

> "After working out our preoperative data we come to the conclusion that there is no connexion between hearing a systolic murmur and mitral insufficiency. We expect to be able to demonstrate with echo- phono- and angiographical investigations, that, also in postoperative patients, the systolic murmur in these patients does not necessarily point to mitral insufficiency."

Puzzled by the origin of the murmur, we initiated a study to investigate all patients that had undergone operation for closure of their "ostium primum defect". We located all such patients undergoing surgery in the 20 year period since 1962, although tracking down some of the patients posed some problems. One patient was traced back to the jungles of inner Congo, from whence we, surprisingly, received an "all is well" accompanied with a chest X-ray and an electrocardiogram. The data from this follow-up study were eventually joined with the data on follow-up of the patients with "complete AV canal". Together these studies form the basis of Chapter IV[10].

Angiography

Because of our interest in the origin of the systolic murmur, which we believed not to be caused by left atrioventricular valvar regurgitation, we started reviewing the preoperative left ventricular angiocardiograms. We were prompted to do so after our attention was drawn to a similarity between this murmur and the murmur often encountered in patients with hypertrophic obstructive cardiomyopathy[11]. These patients often have murmurs originating from stenosis within the left ventricular outflow tract, caused by the hypertrophic ventricular septum. This murmur might not be caused be the stenosis in itself, but by the velocity of constriction of the outflow tract, thus having an impact upon the blood flow which, in turn, might cause the murmur. To investigate this pathophysiologic possibility we reviewed all preoperative angiograms of all "primum defects".

In the right anterior oblique (or antero-posterior in some patients) projection, we measured the diameter of the left ventricular outflow tract in each consecutive systolic frame. After discarding films that were not "measurable" because of extrasystoles, we ended with usable angiograms from 15 patients. From these data it was, in principle, possible to calculate the "velocity" of narrowing of the outflow tract [or the function: d(diameter)/d(time)]. There appeared, however, to be no computer program capable of analyzing on the type of data we had obtained. The problem was that all patients, having different cardiac rates, had a different number of angiographic frames and, thus, a different number of observations per systole. To calculate the mean velocity of narrowing for a group of data with different time intervals, a computer program was written in the language Pascal-6000 (Listing of the CUMCURV program in the appendix). In this manner, we found a correlation between the initial velocity of narrowing and the minimal relative diameter of the left ventricular outflow tract.

Echocardiography

Expanding the study at the same time, however, we accumulated a much larger amount of largely overlapping echocardiographical data (Chapter V). The echocardiographic findings had a more definite clinical context, so we did not pursue our angiocardiographic investigations. All patients that were operated for their "ostium primum defect" that could, and were willing to come, were investigated echocardiographically. Additionally, all patients were auscultated independently by someone who was unaware of the results of echocardiography. Phonocardiography was not done, because quantification of the loudness of the murmur was felt to be impossible. Angiocardiography was not performed because catheterization was not thought to be warranted in a symptomless population.
The first paper to emerge from this research constitutes Chapter V[12]. Unfortunately we were never able to link the murmur to the static or dynamic properties of the left ventricular outflow tract. But we did demonstrate that the outflow tract is long and narrow and that it constricts during systole. The constriction phenomenon was later confirmed by Ottenkamp and Guyt from Leiden[13,14]. Using the CUMCURV computer program (Appendix), we calculated that the velocity of narrowing peaked early in systole in those patients with a narrow outflow tract. Conceivably that phenomenon might be related to the murmur that very often is demonstrably unrelated to left atrioventricular valve regurgitation. When we tried to interpret our echocardiographic findings, however, we ran into anatomical problems, because confusion arose as to the precise anatomical nature of the outflow tract.

The left ventricular outflow tract

The anatomical delineation of the left ventricular outflow tract in connexion with its obstruction then became the subject of a study that was conducted at the Cardiothoracic Institute, Brompton Hospital, London. For reasons of logical cohesion, the results of that study are the subject of Chapter VI[15], although the echocardiographic work on the left atrioventricular valve was integrated in the study on the outflow tract.
The length of the outflow tract proved to depend upon the extent of bridging of the superior bridging leaflet and upon its connexion to the septal crest. Furthermore, we found that the posterior wall of the outflow tract consists of fibrous tissue and atrial muscle which together are analogous to the ventriculo-infundibular fold on the right side of the heart. We designated this structure the "atrial fold". Surgically, it can be resected during replacement of the left atrioventricular valve as described in Chapter VI. Since then, we have performed this manoeuvre a number of times during operation.

The left atrioventricular valve

The results of the echocardiographic study pertaining to the left atrioventricular valve are the subject of Chapter VII[16]. Expanding the notion of a trifoliate left atrioventricular valve, we linked postoperative function to its morphologic disposition. In principle, this would make it possible to determine the preferable operative technique from the observed morphology, the latter either being determined preoperatively by echo or intraoperatively by direct measurements. A crucial table that was omitted at publication (at the request of the Journal editor), is included at the end of the chapter.

Morphometry

The anatomic variability found in the echocardiographic study was finally confirmed at the morphometric study that we performed at the Cardiopathological Museum of the Children's Hospital of Pittsburgh, studying 151 examples of atrioventricular septal defect (Chapter VIII)[17]. We found that the hearts could be analyzed in terms of a sequence of diminishing developmental perfection of the commissures of the atrioventricular valve. When septal and valvar morphology were linked, we were, finally, lead to some embryological speculations. The resultant expanded understanding of the anatomy of atrioventricular septal defect may greatly enhance surgical treatment.

Relevance

Some thoughts on atrioventricular septal defect are collated in Chapter IX, in the framework of an editorial note[18]. One of the salient points addressed is the stance towards children with Down's syndrome and atrioventricular septal defects. Briefly, our hospital's policy for the past decades has been that all children should undergo operation since, if operation is not performed, they are likely to die of the sequelae of Eisenmenger's complex. Furthermore, Down's syndrome is not an incremental risk factor for operative death[19]. And children with Down's syndrome do not have a greater risk of developing elevated pulmonary vascular resistance, as has been postulated in the past[20]. Even if life expectancy was not increased by surgery, a view that I contest, I think that avoidance of the sequels of the Eisenmenger's complex is more than reason enough to offer these patients an operation, this being the same treatment as offered all other children with atrioventricular septal defect.

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CHAPTER II

EVOLUTION OF THE ANATOMICAL PERCEPTION

Few other congenital cardiac anomalies are, and have been known, by so many different names as atrioventricular septal defect. Dr.Thomas Bevill Peacock, physician to the Royal Free Hospital and Royal General Dispensary, Finsbury Circus in London (Fig. 1), was probably one of the first accurately to recognize and describe this anomaly in 1846[1]. On the 7th of December of that year he described a heart of an eleven year old girl "whose history during life was not known". In his remarkably concise and precise account Dr.Peacock describes:

> "The interesting points in the case were, first, a deficiency of the base of the inter-auricular septum, with a perfect closure of the foramen ovale. The deficient space allowed of free communication between the two auricles, so that there could only be said to be one auriculo-ventricular aperture. 2ndly. A distinctly tricuspid form of the left auriculoventricular valve; and 3rdly, a deficiency at the base of the septum of the ventricles, nearly closed by an extension of the anterior fold of the left auriculo-ventricular valve".

THOMAS B. PEACOCK (1812-1882)

Fig. 1.

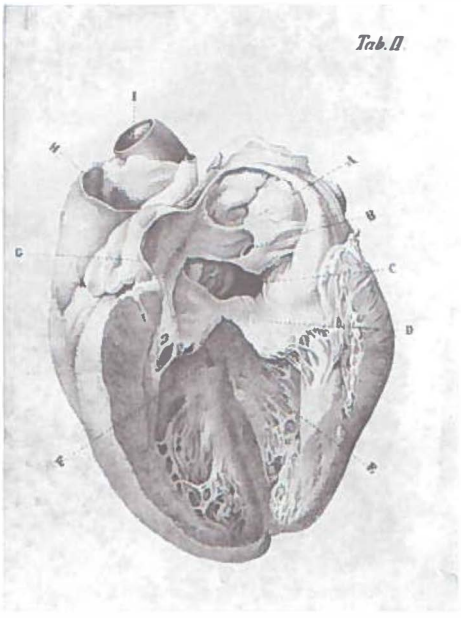

Fig. 2.

Earlier accounts of atrioventricular septal defects are known, notably in the thesis of Alexander Ecker of Freiburg[2], but his description is not as clear concerning the anatomy as is that of Peacock. Ecker, did not recognize the anatomy to be so fundamentally different from normal. In Ecker's thesis, the beautiful drawing of his case 6 is probably one of the finest early pictures of the lesion (Fig. 2). Astoundingly, Peacock's tale is not only one of the first accurate descriptions of an atrioventricular septal defect, but it remained the sole truly correct description for more than a century. Unfortunately, Dr. Peacock's account was largely forgotten. It would last 29 years before the next meticulous description of such a case appeared in Professor Dr. Carl Freiherr von Rokitansky's "die Defecte der Scheidewände des Herzens", published in Vienna in 1875 [53]. The description by Peacock is not referred to by Rokitansky, so he was apparently unaware of this account, although he does refer to Peacock's 1866 book "Malformations of the Human Heart" in connexion with various other congenital anomalies. Rokitansky, however, did not realize or accept that the left atrioventricular valve could be anything but bicuspid:

> "Der Aortenzipfel der Bicuspidalis sei gespalten, wobei sein vorderer Theil mit der Pars membran. septi und dem vorderen Zipfel Tricuspidalis zusammenfliesst und den Zugang zur Aorta von hinten deckt, der hintere mit dem inneren Zipfel der Tricusp. über dem Septum ventr. zusammenfliesst".

"The aortic leaflet of the bicuspid valve was cleft, while the anterior part was connected to the membranous septum and the anterior leaflet of the tricuspid valve; it covered the entrance to the aorta on the posterior aspect. The posterior part was connected to the inner leaflet of the tricuspid valve covering the ventricular septum".

Unfortunately Dr. Rokitansky's descriptions were widely distributed and repeated, while Dr. Peacock's account slumbered in oblivion. It has always been fashionable to call the left atrioventricular valve of the normal heart the "mitral valve" because of its resemblance to a bishop's bicuspid hood. And, when it was forgotten that "mitral" stands for bicuspid, the "cleft anterior leaflet of the mitral valve" was born. This description, nonetheless, negates the true character of the anomaly, and is an anatomical contradiction in terms.

Rokitansky's descriptions were also unfortunate because of his use of the concept of an "ostium primum defect". Peacock wrote: "...deficiency of the base of the inter-auricular septum...", which accurately described the functional anatomical situation. Rokitansky, however, interpreted this as a defect in the septum primum (an embryological structure) that, in the normal heart is not discernable as a separate anatomical entity. Ironically, in this lesion, the primary part of the atrial septum itself is not deficient at all, but present, with its leading edge bordering the defect. The arrangement of the *ostium* primum is debatable, since it is questionable if this communication in atrioventricular septal defect is comparable to that in the normal heart. The misconception of Rokitansky, and others for more than a century after him, is that the atrioventricular valves in hearts with deficient atrioventricular septation can be interpreted in the light of the normal atrioventricular junction. In reality, a so-called atrial septal defect of the "ostium primum type" is a defect that functions as an interatrial communication simply because of the apical displacement of the atrioventricular valves, the atrial septum itself being developmentally intact.

Persistent "atrioventricular canal" or "ostium atrioventriculare commune" is another embryologically based term, used by Monckeberg in 1923 and by Maude Abbott in 1936[4,5]. In the embryo, the canal is a well-recognized structure. But, in normal or malformed hearts, this structure is not separately recognizable. Furthermore, although there is certainly a common atrioventricular junction in the heart under discussion, this relates primarily to the lack of any septal junction rather than to persistence of a truly embryologic atrioventricular canal. "Endocardial cushion defect" is a term minted by two thoracic surgeons of the Harvard Medical School, Watkins and Gross, in 1955[6] and endorsed by Van Mierop in preference of "atrioventricular canal" and "persistent ostium primum"[7].

Arthur Keith, nonetheless, suggested as early as 1909 in his Hunterian lectures on malformations of the heart, that non-fusion of the endocardial cushions pro-

duced this anomaly, analogous to the pathogenesis of a hare lip[8]. Watkins and Gross justified their choice by:

> "We prefer to classify this defect as one of the endocardial cushion because it is related not only to inadequate fusion of the primitive atrial septum with the endocardial cushions (the anlage of the mitral and tricuspid valve rings) but also the incorrect maturation of the endocardial cushions themselves. This maldevelopment results in serious atrioventricular valve deformity and regurgitation."

Again it is the implicit embryologic basis of "endocardial cushion defect" that provides the fuel for the future debates. Recent evidence indicates that the endocardial cushions contribute very little to the atrioventricular valves and therefore the embryological basis for this name has vanished[9,10], although future research may well demonstrate that failure of fusion of the two cushions does play a role in the development of atrioventricular septal defects.

Why cannot we just describe what we see, without interpreting the facts in the context of the theories that are fashionable at the time?

Particularly in atrioventricular septal defect, it seems difficult to refrain from the use of nondescriptive interpretative nomenclature. Apparently, on the other hand, the complexity of the anomaly (involving not only the ventricular and the atrial septum and the atrioventricular valves, but also the general cardiac architecture) defies a concise anatomic description. While, on the other hand, an embryologically based description seems to have the advantage of being short and smart if the entire anomaly could be traced to the maldevelopment of one known embryologic structure.

Alas, this has not proven to be the case. "Atrioventricular septal defect" is the best of all proposed names until now, because it does not implicitly claim facts that cannot be substantiated[11]. On the other hand, a definite disadvantage of this name is that the information given is incomplete. Not only the atrioventricular septum is deficient, but a variety of other cardiac structures are grossly deformed, for which several suffixes should be added concerning the partitioning of the atrioventricular ostium and the potential abnormal communications between the different cardiac chambers.

Furthermore, rare hearts exist with a communication between the right atrium and the left ventricle due to deficiency of the normal membranous component of the atrioventricular septum. Those membranous atrioventricular septal defects, nonetheless, have none of the other anatomic stigmata of the hearts discussed in this thesis. Is there an alternative title? In analogy to the ventricular septal defect, with an overriding aorta and with infundibular and valvar pulmonary stenosis, which usually goes by the name of Fallot, it might have been better to call the anomaly of this study:

PEACOCK'S DISEASE

The use of this name has the advantage that it claims nothing but the alleged first accurate description by Dr.Thomas Bevill Peacock. It precludes embryologic discussions outside a true embryologic context and it has the advantage that we can focus on the surgical morphology without compromising ourselves with outdated theories. But, sadly, calling disease by investigator's names is not fashionable. I shall stick, therefore, with the title "atrioventricular septal defect".

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CHAPTER III

SURGICAL EVOLUTION

Surgery of atrial septal defects was probably started by Gordon Murray of Toronto, Canada[1]. In his article of 1948, he described first three cases of ventricular septal defect. He tried to close these with the use of strips of fascia lata 3/4 of an inch wide, which he blindly threaded across the ventricular septal defect. Total closure of the defect was probably not achieved by this method, although he described no mortality, and the patients appeared to have improved after surgery. In his fourth case, however, the atrial septal defect was closed by silk sutures:

> "It was decided, therefore, to place our sutures of silk, in this case, beginning to the right of the aorta and pulmonary artery and emerging posteriorly through the area between the superior vena cava and the right pulmonary veins. Two sutures were passed through without difficulty. (It is the plan in future cases to pass more than two sutures.) These sutures were tied together posteriorly and were drawn taut from the anterior end and compressed with the finger. This caused a very great change in the size, shape and color of the heart. The right auricle diminished to at least one-half its size within about two minutes. The patient's general condition was good. The blood pressure remained normal. The sutures, after experimenting with this for some time, were tied down firmly, compressing the anterior and posterior walls of the auricles. This caused continued improvement in the patient's condition. The chest cavity was then closed without drainage."

Further information on this patient, derived from the paper by Keith and Forsyth[2], disclosed that at subsequent cardiac catheterization, 14 months postoperatively, there still was a substantial left-to-right shunt and significant right heart hypertension. The patient had at least one episode of cardiac failure during this period. Cardiac X-ray showed some generalized decrease in size. Murray's method was, therefore, not employed by anyone else, as far as is known. Henry Swan developed, experimentally, a method in which both auricles were invaginated and sutured on plastic buttons more or less inside the atrial septal defect[3]. He first applied this in a human in October, 1949. Swan's method was clinically employed by Charles Bailey, but the results were very poor, six out of nine patients dying[4]. Four of these were attributed to Lutembacher's syndrome (atrial septal defect combined with mitral stenosis). In the tenth operation, Bailey used his novel method or atrio-septo-pexy (11 Jan. 1952). In this technique, he sutured the redundant right atrial free wall to the margins of the atrial septal defect

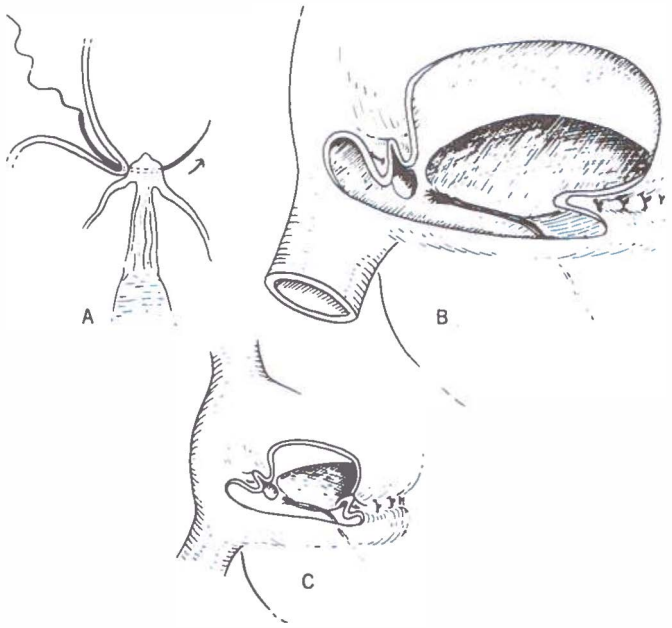

Fig. 1. Bailey's atrioseptopexy.

under the guidance of a bare finger, inserting the suture through the right atrial appendage. On 22 May 1952, Bailey used this technique to close an "ostium primum defect" in the Hahnemann Hospital in Philadelphia, Pa. USA[5]. This is probably the first atrioventricular septal defect ever to undergo surgical closure. In order to close the lowermost part of the defect, Bailey used Murray's technique: "one or more anterior- posterior approximating mattress sutures". The patient was reported to have recovered well. Before operation Bailey, used to inspect visually the defect[6] by use of the Bolton cardioscope[7] (Fig.). But there would be no great future for this technique for the treatment of "ostium primum defect", because Bailey reported a mortality of 11 patients out of 16 (69%) in 1954[8]. This high mortality was due to surgically induced heart block and the inability in this technique to deal with the left atrioventricular valve, so that severe valvar regurgitation often developed or persisted after the operation. In 1955 Bailey stated, while discussing a paper by Watkins and Gross, that he had stopped operating on "septum primum" cases because of this unacceptably high mortality[9].

The paper that Bailey discussed dealt with the results of surgical repair of atrial septal defects using the "atrial well technique" which Robert Gross and Elton Watkins, of the Harvard Medical School, had first described in 1953[10,11]. Gross, incidentally, was also the first one to describe the successful closure of a patent ductus arteriosus in 1939[12]. In the atrial well technique, these surgeons employed

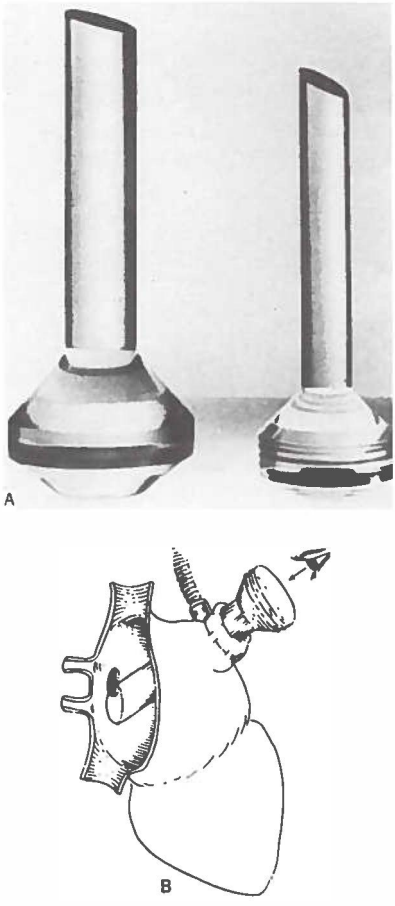

Fig. 2. The Bolton cardioscope.

a rubber funnel-like well, which was sutured onto the atrial appendage, and which subsequently spontaneously filled with blood by the venous pressure. Through this "well", it was possible to perform intra-atrial surgery, albeit blindly. But again this blind technique did not result in an acceptable survival as they described in 1955:

> "We have seen this defect 4 times in the 43 cases. Each time it has been associated with deformities of both the mitral and tricuspid valves. Although we have been able to close some of these defects surgically, cardiac failure and death have promptly supervened. We question the wisdom of attempting to convert the left atrium into a small closed chamber into which blood regurgitates without any passageway for decompression through the surgically closed defect."

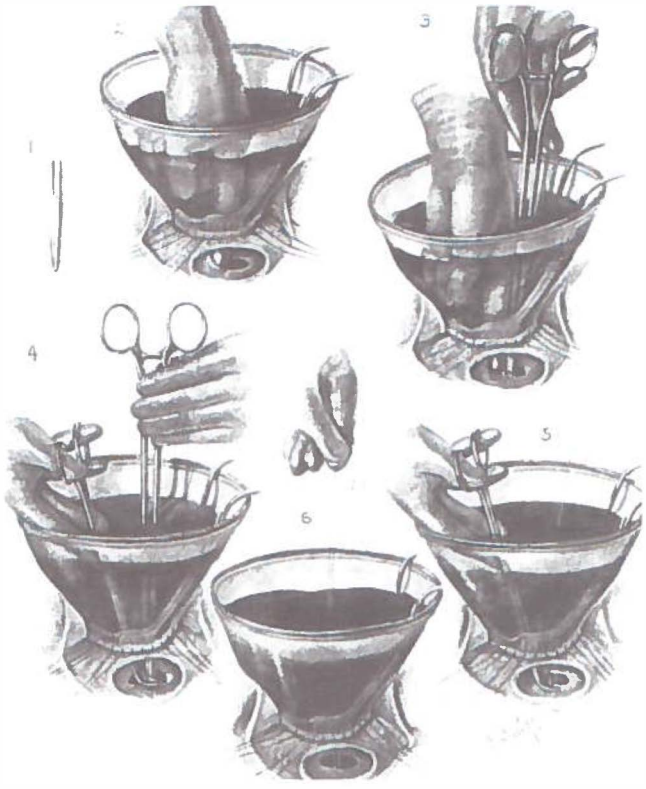

Fig. 3. The atrial well technique.

Very few reports have been made of successful employment of this technique for the repair of "ostium primum defect", again because of the inability to deal with the regurgitant left valve and the risk of damage to the conduction system. The earliest reported successful correction for "common atrioventricular canal" probably took place on January 6, 1954 and was performed by John Kirklin and coworkers at the Mayo Clinic, utilizing the atrial well of Gross[13]. Graciously, but incorrectly, Kirklin gives the credit for the first operation of "atrioventricularis communis" to Lillehei, who was the first to use an open technique (on 26 March 1954), with direct vision, utilizing cross-circulation with a volunteer (usually a parent)[14].

This technique permitted occlusion of the caval veins for correction of the anomaly under direct vision, while brief occlusion of the aorta could make the operating field totally bloodless. Two atrioventricular septal defects were corrected in this way, one dying of "intolerable stenosis of the aortic outflow tract."

"...the defect was approached through a right atrial cardiotomy. Exposure of the defect was excellent. The upper margin of the ventricular septum was sutured to the lower edge of the atrial septal defect with interrupted silk, the stitches being placed through the origin of the common mitral- tricuspid valve. Because of the somewhat more complicated nature of this lesion, the intracardiac time required to effect this repair was twelve and one-fourth minutes. The patient has been completely well clinically since surgical correction."

But the use of Gibbon's pump oxygenater, enabling open cardiotomy, was already supplanting most other techniques, even in 1955[15].

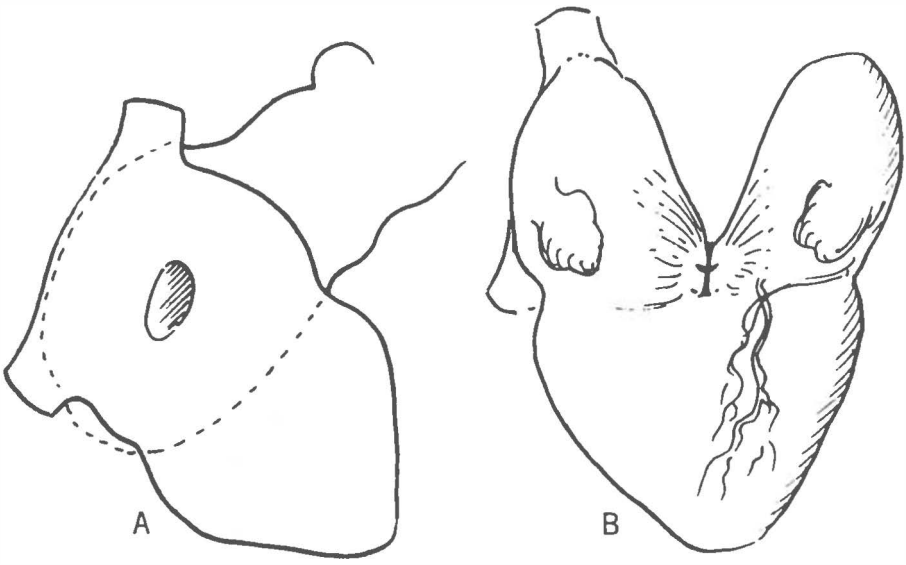

Fig. 4. Søndergaard's circumclusion.

Another closed technique that deserves mention is the circumclusion technique, developed experimentally by Søndergaard in November 1950[16], and then used clinically by Björk in 1953[17], and by Søndergaard himself. By means of a probe, a suture is passed through the lower margin of the defect, starting posteriorly between the superior caval vein and the right lung veins and emerging anteriorly out of the atrial septum. Surprisingly two of the six cases are described as "ostium primum cases", which seems miraculous to me, because it hardly seems possible to pass a probe through the very narrow edge of tissue at the crest of the ventricular septum where the two atrioventricular valves meet. One of the pa-

tients was left with a residual shunt though. Although Björk had reportedly used this technique a few times for a "secundum defect", circumclusion had not caught on and Søndergaard himself had apparently abandoned circumclusion for "primum defects" as his series reported in 1980 makes no mention of this early experience[18]. But he appears still to use it successfully in "secundum defects" as he reported in 1984[19].

The advent of hypothermia offered, for the first time, the possibility to visualize what one was doing in intra-cardiac surgery. Research on hypothermia was, among others, done by Bigelow[20], Boerema[21] and Homan van der Heide[22], with their respective colleagues. Hypothermia for the operation of an atrial septal defect was reportedly first used in 1954 by Lewis, Varco and Taufic, of the University of Minnesota Medical School[23]. Although their repair of "secundum defects" was successful, the limited amount of available intra-cardiac operating time deemed it impracticable for the repair of "primum defects". Their third patient had a "primum defect" and died of a heart block. But, after their innovative work, hypothermia has for a long time remained the technique of choice for the repair of uncomplicated atrial septal defects.

The clinical development of extracorporeal circulation in 1955 eventually brought the revolution that also made repair of "ostium primum defect" feasible through the availability of intra-cardiac operating time, essential for the careful

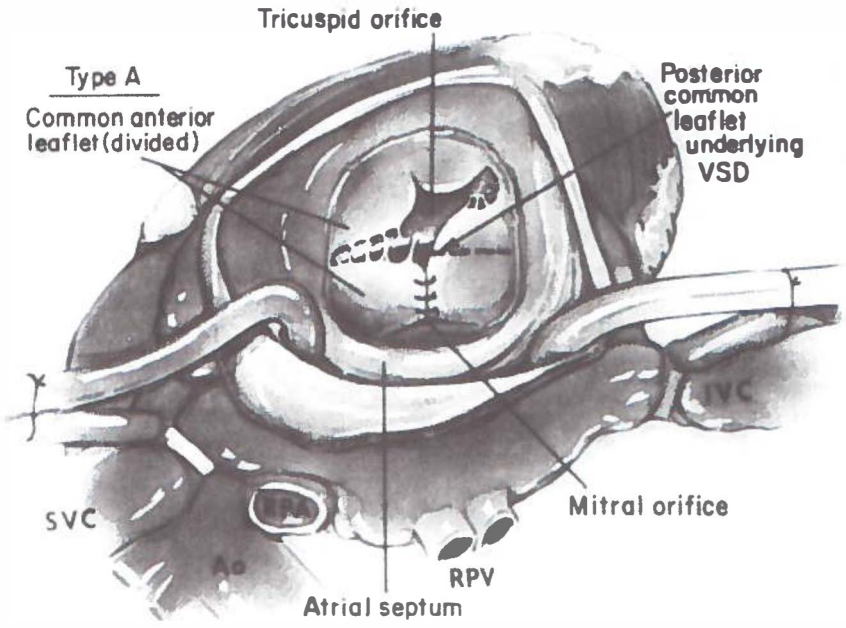

Fig. 5. The Mayo clinic's repair.

management of atrioventricular septal defects. One year later, in 1956, Jack Cooley and John Kirklin of the Mayo Clinic reported the first good results in atrioventricular septal defects with and without interventricular communications[24]. The employment of a pump-oxygenator and extra-corporeal circulation provided the necessary time and vision and was undoubtedly instrumental in these superior results. Twelve patients underwent operation and three died, two because of atrioventricular regurgitation because no attempt was made to correct the incompetence. Undoubtedly, the use of extra-corporeal circulation was the revolution that made surgery of these defects feasible with an acceptable risk. The credit for this development goes to all those who worked on the heart-lung machine, most notably Gibbon[25], who had been working on this since the thirties[26]. But a number of Dutch scientists were also working on the heart lung machine, such as Jongbloed[27], Dorlas[28] and Homan van der Heide[29]. As a result of these efforts the first open heart surgery in the Netherlands was performed in Groningen on 8 May 1957, by Prof.Dr. L.D. Eerland and Prof.Dr. J.N. Homan van der Heide.

A major contribution to surgery was made by the elucidation of the surgical anatomy of the conduction system in atrioventricular septal defect by Lev in the fifties[30]. Since practicable cardiac pacemakers were still decades away, surgically created total heart block was lethal at that time. Since 1956, numerous series with quite acceptable results have been published[31-40]. Much has changed over 30 years in the results of primary repair. Carpentier has focused on the left atrioventricular valve and the way in which it should be approached[41]. His great feat is the

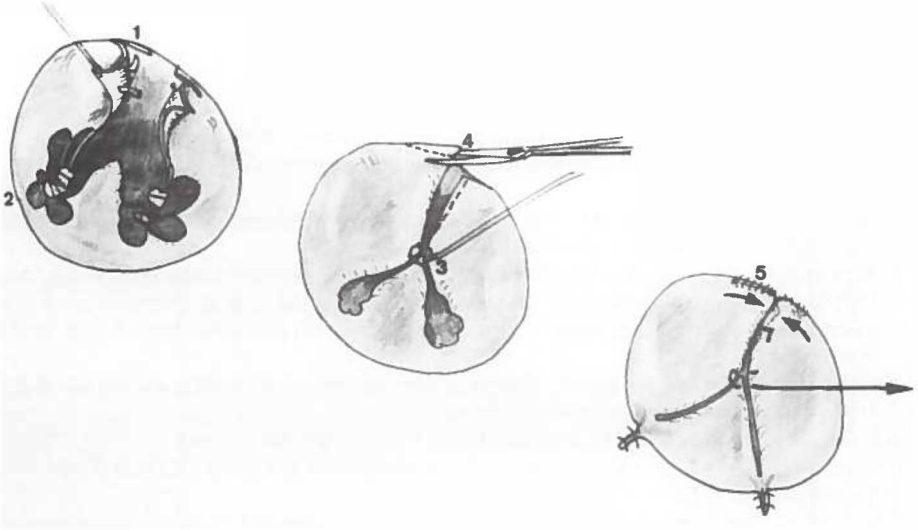

Fig. 6. Carpentier's trifoliate repair.

surgical management of the left atrioventricular valve based on the trifoliate concept. For more than a century since Peacock, surgeons and morphologists have considered the anterior leaflet of the mitral valve to be "cleft", and thus something to be closed. The discussion has been: "to close or not to close the cleft", disregarding the more complex nature of the anomaly.
Those who always closed the cleft probably lost patients due to stenosis of the valve. Those who never closed it lost a different category of patients to regurgitation. Carpentier and Penkoske[42] have pointed out that the left atrioventricular valve in this anomaly does not resemble morphologically a mitral valve. It is not only anatomically impossible to make the valve "mitral" or bicuspid, neither is it surgically advisable. When one accepts the concept of a regurgitant trifoliate left atrioventricular valve, that should be repaired, but will stay trifoliate, then the surgical results will improve.
Major contributions have been made concerning the precise delineation of the conduction tissue, and its subsequent surgical avoidance. Thiene et al.[43] and Kurosawa and Becker[44] have published detailed studies. In order to avoid damage to the conduction system, Kirklin and Barratt-Boyes advocate leaving the coronary sinus on the left side of the patch[45], but this does not always seem to be necessary[46]. In this thesis, nonetheless, I will not comment on the conduction system further. Instead, I will concentrate on the behaviour of the left ventricular outflow tract, its surgical anatomy (especially in relation to replacement of the left atrioventricular valve), and with the morphological variability of the left valve and the relation to its postoperative function.

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CHAPTER IV

LONG TERM FOLLOW UP OF 133 ATRIOVENTRICULAR SEPTAL DEFECTS.

Tjark Ebels, Adri H.Cromme - Dijkhuis, Anton Eijgelaar, Miek J.M.Schasfoort - van Leeuwen, Erik J.Meijboom, J.N.Homan van der Heide.

Abstract

From 1962 through 1986 we operated 133 patients with atrioventricular (AV) septal defect (SD). Median follow-up time is 8 years. Ninety of these 133 had separate AV orifices and 43 had a common AV orifice. Primary complete repair was done in 129, pulmonary artery banding was done in 3, of which 2 underwent subsequent repair, and one patient with a concomitant severe pulmonary stenosis received a shunt. Total (operative and late) mortality was 23 patients (17%, 70% Confidence Limits (CL) 14% - 22%). Left AV valve replacement was done in 8 patients (6%; CL: 4% - 9%), never at the primary correction; of these, 4 patients died (50%; CL: 27% - 73%).
Logistic regression analysis showed that 1. common AV orifice, 2. inability to adequately repair the left AV valve, and 3. major associated anomalies to be incremental risk factors for death. Late follow up was obtained in all patients. Echo-doppler investigation -more than one year after correction- of the left AV valve was obtained in 78 of the 110 survivors.
The doppler severity of regurgitation was semi-quantified and the Left Atrium (LA) / Aorta (Ao) ratio was measured. None or little regurgitation was found in 66 patients (85%; CL: 79% - 89%), moderate regurgitation in 10 patients (13%; CL: 9% - 18%), while severe regurgitation was found in only 2 patients (3%; CL: 1% - 6%). There was no difference in regurgitation or LA/Ao ratio between patients with common or separate AV orifices. Our conclusion is that most survivors show some regurgitation but LA/Ao ratio usually is in the upper range of normal. Most patients are able to lead a normal life without physical limitations. Residual severe left AV valve regurgitation remains a risk factor, that should be avoided at all costs. This is even more compelling because of the high mortality associated with late left AV valve replacement.

Introduction

Corrective operation for atrioventricular (AV) septal defect has become a routine procedure with low morbidity and mortality, depending on the specific anatomy[1,2,3]. The success of the operation seems to depend largely on the functional result of left AV valve repair. That this valve is in effect trifoliate and, therefore, not mitral or bicuspid (as a bishop's hood) has been a major rediscovery of the last decade by Carpentier[4], after its first accurate description by Peacock in 1846[5].

The determinants of long term follow-up though, are, by their very nature, only gradually beginning to emerge. The function of the left atrioventricular valve, again, seems to have a paramount influence on the wellbeing of the patient. To determine the short and long term results and the factors that influence them, we analyzed all the patients that were operated upon for AV septal defect at our institution.

Materials and methods

From 1962 to 1986 we operated 133 patients with AV septal defect. Excluded were patients with concomitant double outlet ventricle and discordant connexions, because we feel that these anomalies dominate the nature of the repair. Included were patients with ductus arteriosus, atrial septal defect, aortic coarctation, cor triatriatum, pulmonary stenosis, and atrial isomerism (Table 1). Ninety of these 133 had separate AV orifices (68%; 70% Confidence Limits (CL):

TABLE 1.

Major associated anomalies

Anomaly	number	
Isomerism Total		4
left	3	
right	1	
Severe pulmonary stenosis		4
Cor Triatriatum		3
Hypoplastic ventricle		2
Absent ventricular septum		1
Total		14

63% - 72%), which used to be called "ostium primum defects" or "partial AV canal". Common AV orifice was found in 43 patients (32%; CL: 28% - 37%), which used to be called "complete AV canal". All survivors were recalled for echocardiographic and doppler evaluation, specifically of their left AV valve.
Simple contingency tables and chi-square tests were used for individual variables and their relations. Multivariate logistic regression analysis with tests of significance were made for the analysis of incremental risk factors for death. All p-values less than 0,05 we considered to indicate significant relations, those of 0.05 to 0.1 probably true ones, and those of 0.1 to 0.2 possibly true ones. Variables with a p-value greater than 0.2 were rejected. Failure of left AV valve repair was defined as either moderate or severe regurgitation at postoperative evaluation at any time, or replacement of the valve by a prosthesis.

Results

Of the 133 patients 74 (56%; CL: 51% - 60%) were female and 59 (44%; CL: 40% - 49%) were male. Of the 43 patients with common AV orifice 27 (63%; CL: 54% - 71%) were female and 16 (37%; CL: 29% - 46%) were male. So girls seem to be more likely to have a complete AV septal defect than boys, but this result is just possibly significant (p=0.1273; 2-tailed binomial test). This is independent of the presence of Down syndrome, which had an equal sex distribution.
Of the 133 patients 129 (98%; CL: 96% - 99%) underwent primary complete correction at which the intracardiac shunt was eliminated by one or two patches, and the left AV valve was repaired. Three patients (2%; CL:1%- 4%) underwent primary pulmonary artery banding, of which two underwent secondary complete repair. One of these three patients developed a very high pulmonary vascular resistance despite the banding and is now considered to be inoperable. Another patient with severe pulmonary stenosis underwent an aorto-pulmonary artery shunt operation, and died.
Median follow up of all patients was 8 years, range 1 - 25 years. Total mortality (operative and late) was 23 patients (17%; CL: 14% - 22%). The total mortality was 4.5 times as high in the group of patients with common AV orifice as in those with separate AV orifices (Table 2).
Actuarial analysis showed that at 25 years survival was 90% (SEM[1]: 3.6%) for patients with separate AV orifices and 61% (SEM: 7.6%) for those with a common AV orifice (Fig. 1). The mortality in those patients with a common AV orifice fell entirely within 30 days after operation, and was largely connected with failure of left AV valve repair.

[1] SEM: Standard Error of Mean

TABLE 2.

Total mortality after operation for atrioventricular septal defect

TYPE	NUMBER	MORTALITY	%	CL
COMMON	42	16	38.1	30 - 47
SEPARATE	90	7	7.8	5 - 12
TOTAL	132	23	17.4	14 - 22

Legend: CL: 70% Confidence Limits.
Mortality: Operative as well as late mortality.

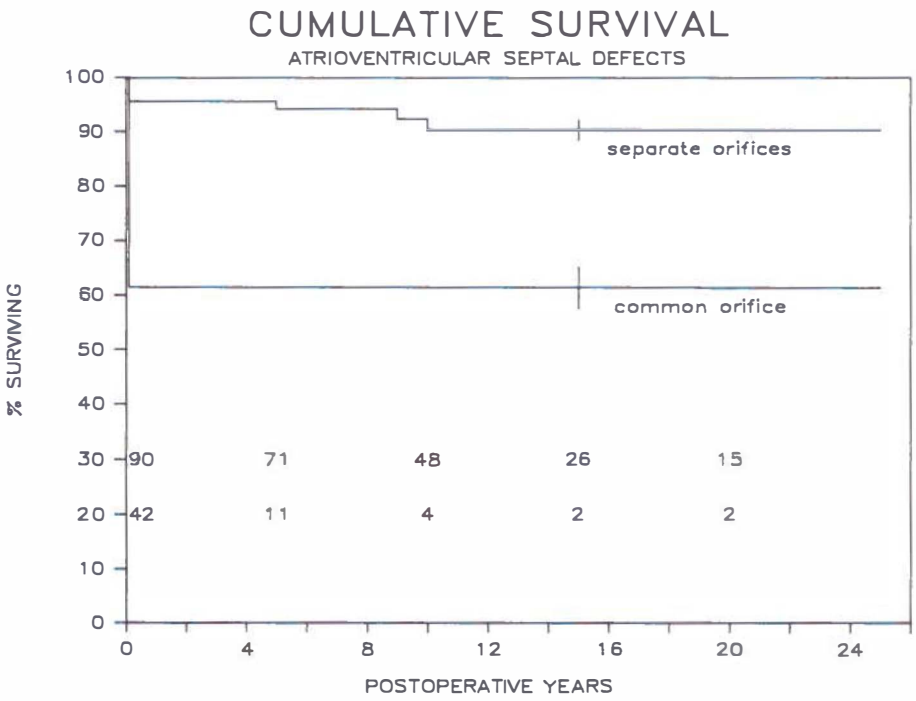

Fig. 1. Actuarial analysis of the cumulative survival of patients with Atrioventricular septal defect. The numbers whithin the graph signify the number of patients at risk at a certain period of folow up.

In contrast, operative mortality in patients with separate AV valve orifices was connected with major associated anomalies. Late mortality, however, in these patients was associated with late AV valve replacement.
Down syndrome was found in 30 patients (23%; CL: 19% - 27%). Common AV orifice was 9 times as frequent as separate AV orifices in children with Down syndrome. In children without Down syndrome the distribution is reversed: separate AV orifices was 5 times as frequent as common AV orifice (Table 3). Down syndrome did not have a separate influence on mortality.

TABLE 3.

Breakdown of all AV septal defects according to Down syndrome and morphology of the AV orifice.

TYPE	DOWN SYNDROME		
	NO	YES	TOTAL
COMMON	16	27	43
SEPARATE	87	3	90
TOTAL	103	30	133

Chi-square: 55.5; $p = 0.0001$

But in the late seventies and early eighties we changed our policy, and started operating upon all children with common AV orifice with or without Down syndrome, preferably before the age of one year. Initially operative mortality increased as the result of this policy, because of a learning curve, and sometimes too high pulmonary vascular resistance. Mortality dropped again towards the end of the study, and was 2 out of 8 patients (25%; CL: 9% - 50%) in 1986; these fatalities were caused by endocarditis and a hypoplastic right ventricle.
Left AV valve replacement was done in 8 patients (6%; CL: 4% - 9%) and was never done at the primary correction. The mean interval between correction and valve replacement was 3.3 years (range 1 day - 8.2 years). Three valve replacements were done in children with a common AV orifice (7%; CL: 3% - 14%) and five were done in the setting of separate AV orifices (6%; CL: 3% - 9%), so there was no difference in incidence. Four of these eight patients died (50%; CL: 27% - 73%). One patient died when valve replacement was done shortly after primary correction; one died late of rhythm disturbances, and one died through

TABLE 4.

Regurgitation of the left AV valve in AV septal defect. Echo-Doppler measurements.

REGURG.	ORIFICE		
	SEPARATE	COMMON	TOTAL
NONE	24	8	32
LITTLE	24	10	34
MODERATE	6	4	10
SEVERE	0	2	2
TOTAL	54	24	78

Legend: Regurg.: regurgitation Chi-square test: 5.43. No significant difference.

TABLE 5.

Incremental risk factors for death.
(23 deaths among 133 patients)

Incremental risk factor	Logistic coefficient ± SE	p-value
Presence of common AV orifice	1.00 ± 0.52	0.0428
Left AV valve repair failure	1.23 ± 0.34	0.0001
Major associated anomaly	2.04 ± 0.50	0.0001
Year of first operation	-0.003 ± 0.076	0.0546
Age (months)	0.094 ± 0.055	0.1380
Interaction of age with year of operation	0.0014 ± 0.0008	0.0520

Intercept = 0.95 ± 6.03
Legend: SE: Standard Error. AV: Atrioventricular
Major associated anomalies include: isomerism, cor triatriatum, ventricular hypoplasia, severe pulmonary stenosis (Tetralogy of Fallot).

outflow tract obstruction by the prosthesis. Finally, one of these four patients had a virtually absent ventricular septum, which greatly complicated repair.
Seventy-eight of the 109 survivors (72%; CL: 66% - 76%) had an echo- doppler investigation of sufficient quality to permit semi-quantification of left AV valve regurgitation; some refused to cooperate in the study and in some patients the echo quality was not good enough to obtain all details, mainly due to obesity. The severity of regurgitation of the left AV valve was not different between patients with common or separate AV orifices (Table 4). Neither was the mean ratio of left atrium diameter versus aorta diameter on the echocardiogram, being 1.35 in the patients with separate orifices and 1.24 in those with a common AV orifice.
Logistic regression analysis showed three factors to be important incremental risk factors for death. These factors were: 1. common AV orifice, 2. inability to adequately repair the left AV valve, 3. major associated anomalies (Table 5). Year of the initial operation was possibly an incremental risk factor. Over the years common AV orifice became less of a risk factor.

Comment

AV septal defect is an anomaly that can now be operated with relatively low operative mortality[6,7]. The only factors influencing operative mortality are major associated anomalies, the surgical inability to adequately repair the left AV valve, and the presence of a common AV orifice. The usual trifoliate left AV valve should not pose serious surgical problems, although the specific valve morphology might call for individualised surgical technique[8,9]. To be able to predict the morphology and hence the optimal surgical technique is one of the echocardiographical challenges of the future. Certainly some of the morphologic abnormalities are very difficult to manage, such as double orifice or "parachute" left AV valve [10,11]. Failure to produce a competent valve results in serious sequelae. Late valve replacement still carries a high mortality[1], while the prospect of a life with a valve prosthesis is cumbersome, especially for a child. Inadvertent obstruction of the left ventricular outflow tract by the AV valve prosthesis is probably the reason for the high mortality of AV valve replacement. We may have solved this operative problem, though, by resection of the posterior wall of the outflow tract[12]. This procedure is not very frequent, so the impact on the long term results does not show yet. But, fortunately, most of our patients are well and able to lead normal lives without signs of serious valve dysfunction.
Down syndrome has not been a risk factor, and we have been very convinced of our policy to operate on all these patients too. Lately, the issue has been raised whether or not survival is better with operation in these children[13]. We think it

is, although the present study does not permit any conclusions to that extent. But the prospect of a Down syndrome child dying of the sequelae of irreversible pulmonary hypertension is distressing to say the least. Therefore we feel it is warranted to offer these children the chance of a normal life, through a corrective operation.

A word of caution remains about the interpretation of surgical series such as this. It is very evident that the change in policy that we made around 1980 had a large impact on the figures here presented. Obviously similar patients existed before 1980, but they do not appear in this study, since we limit ourselves to surgical patients. Therefore, the policy whether or not to operate, and likewise, whether or not to include in a study has a paramount impact on the figures. Ideally analysis should start as early as possible, specifically including patients not operated upon, which can have a variety of reasons. Actually these last patients can be considered to be more interesting than those operated, but they are seldomly reported, let alone separately analyzed, except in a recent publication by Clapp and associates[14].

Finally, we conclude that AV septal defect can be operated upon with low mortality, also for children with Down syndrome. Adequate repair of the left AV valve is of major importance for the future wellbeing of the patient.

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CHAPTER V

ANATOMIC AND FUNCTIONAL "OBSTRUCTION" OF THE OUTFLOW TRACT IN ATRIOVENTRICULAR SEPTAL DEFECTS WITH SEPARATE VALVE ORIFICES ("OSTIUM PRIMUM ATRIAL SEPTAL DEFECT"): AN ECHOCARDIOGRAPHIC STUDY.

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Summary.

Left ventricular (LV) outflow tract (OT) obstruction can be treacherous in any form of atrioventricular (AV) septal defect. The properties of the LVOT were investigated echocardiographically in 64 patients with separate valve orifices ("ostium primum atrial septal defect") who had survived corrective surgery. M-mode and cross-sectional echocardiographic (echo) images were made of the LVOT. The degree of malalignment of the aorta with the ventricular septum, the left atrium-aortic ratio, the fractional LV shortening and the diameter of the LVOT were recorded. Fixed anatomical obstruction was found in 3 patients, consisting of muscular bands or abnormal attachment of tension apparatus. Malalignment of the aorta with the ventricular septum was found in 62% of the patients. The diameter of the LVOT was smaller than that of the aortic root in 71% of the cases. The mean diameter of the LVOT was 92% ± 27% (range 35% to 143%) of the aortic root diameter. Because its walls are mainly muscular, the LVOT constricts during systole. The mean end-systolic diameter of the LVOT was 77% ± 22% (range 23% to 129%) of the aortic root diameter. Sequential measurements showed that the LVOT constricted gradually, but the velocity of constriction in patients with the most severe narrowing showed a distinct maximum in the first fifth of systole. In conclusion, a series of elements contribute to

a potentially perilous arrangement of the LVOT in patients with AV septal defect. This intrinsically narrow tunnel was constricted during systole by its muscular walls. This potential for LVOT obstruction was seen to be greater when the aorta was closer to the chest wall on the M-mode measurement than was the left side of the septum. Any patient with an AV septal defect should be assessed for the potential of LVOT obstruction. Any degree of fixed stenosis produces immediate overt obstruction. The systolic murmur often attributed in these patients to AV valve regurgitation may also originate from the narrowed subaortic outflow tract.

Introduction.

Left ventricular (LV) outflow tract (OT) obstruction has been described as an insidious entity in atrioventricular (AV) septal defects (endocardial cushion defects or AV canal malformations).[1-3] In these lesions, the LVOT is an abnormally elongated tubular structure made up posteriorly by an area of aortic-AV valve continuity, bordered by the LV free wall, the infundibular septum and the ventriculo-infundibular fold. Piccoli et al, in a study of autopsy hearts, found that the LVOT in cases with separate valve orifices (atrium primum septal defect) was anatomically obstructed to varying degrees.[4] In some cases it was obstructed by abnormal attachment of the valvular tension apparatus and in others the aorta was malaligned with the ventricular septum. Fixed subaortic obstruction producing substantial pressure gradients have been described only rarely in this lesion. However, the frequently encountered systolic murmur that is not due to left AV valve regurgitation has been attributed to stenosis within the LVOT[3,5] To assess the properties of the LVOT, in AV septal defects with separate AV valve orifices, we performed an echocardiographic (echo) study in patients who had successfully undergone corrective surgery. The echo study was designed to assess the potential for functional or anatomic LVOT obstruction.

Methods.

Subjects: We followed 75 patients with AV septal defects with discrete right and left AV valve orifices who underwent operation at the University Hospital in Groningen from 1962 to 1982. The diagnosis of ostium primum atrial septal defect had been established by physical examination, chest radiography (increased cardiothoracic ratio mean 59% ± 8%, range 47% to 76%), electrocardiogram (increased PR interval, mean 178 ± 38 ms, range 100 to 260 ms, and left-axis deviation mean -60° ± 23°, range 0° to -120°) and cardiac catheterization with LV cineangiography (all patients showed a goose neck deformity).

Surgery was performed in a standardized fashion. The atrial septal defect was closed with an autologous pericardium-Teflon® double patch and the "cleft" was closed until it was judged that maximal competence of the left AV valve had been achieved. In several cases abnormal tension apparatus attached within the LVOT was removed. Sometimes the inferior (posterior) leaflet of the left AV valve needed to be mobilized because of abnormal chordal attachments.

Sixty-nine patients (92%) survived until 1983, 64 (85%) of whom could be recalled for the echo study. Echo study did not supply all data in 14 patients because of technical problems. Measurements that were obtained were used in the study.

All patients were studied with an Advanced Technology Laboratory mechanical sector scanner (Mark 500). The patients had complete evaluation of their LVOT, using subxyphoid parasternal (4-chamber) long- and short-axis views and an apical 4-chamber view.[6]

M-mode echo examinations of the ascending aorta, left atrium, left- sided AV valve and left ventricle were recorded by sweeping the M-mode control through a good-quality cross-sectional parasternal long-axis view. The parasternal long-axis view and the slightly angled subxyphoid view were used to assess anatomic subvalvular obstruction resulting from an abnormal position of papillary muscles.

The degree of malalignment of the aorta was visualized in the parasternal long-axis view at the end of systole. Malalignment appeared to be a function of rightward displacement of the aorta and septal thickness. The M-mode sweep obtained in this view was used to calculate the percentage of malalignment using the formula:

[(distance transducer-anterior aortic wall)/(distance transducer-left ventricular septal wall) x 100] (Fig. 1).

Particular care was taken to position the transducer in an intercostal space equidistant to the ascending aorta and ventricular septum above the aortic valve leaflets so as to avoid diagnosing spurious malalignment or non-malalignment. The narrowness of the passage between the area of aortic- AV valve continuity and the septum was assessed.[7-9]

The diameter of both the LVOT and the aortic root were calculated from the same M-mode tracing. We measured sequentially the diameter of the LVOT in order to determine the behaviour of the LVOT during systole (Fig. 1). The sequential diameters of the LVOT were expressed as a percentage of the aortic root diameter and as a percentage of its initial diameter in systole. To obtain the velocity of LVOT narrowing during systole, we calculated the first differential of its diameter against time (CYBER 170/760 computer).

Premature closure of the aortic valve as an indicator of subaortic stenosis was evaluated in the M-mode tracing of the aortic valve.

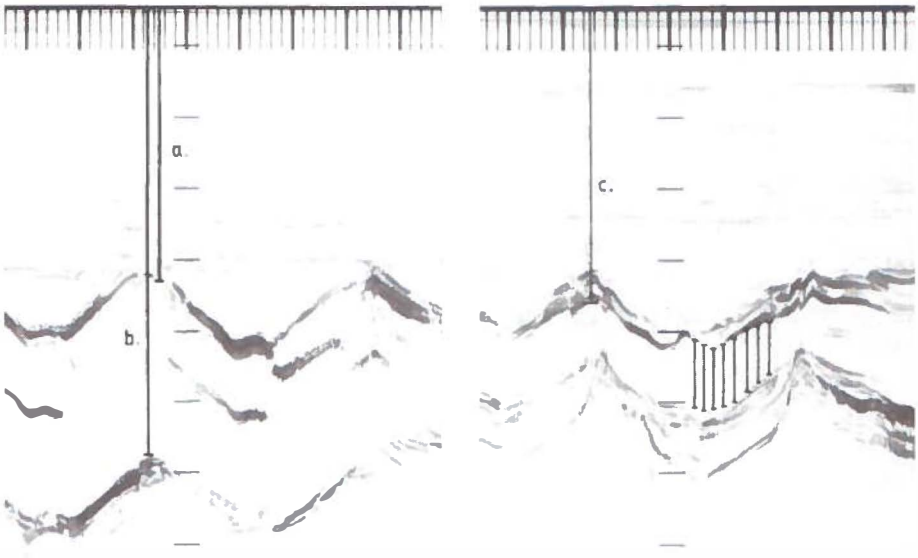

Fig. 1. Measurements of the aorta and the left ventricular outflow tract in the parasternal long-axis view: distances from the transducer to **a**, the anterior aortic wall, **b**, the posterior aortic wall and **c**, the septum. The aortic overriding ratio is calculated as (a/c) x 100. The diameter of the left ventricular outflow tract is measured sequentially, during 1 systole **(left panel).**

Results.

Fixed anatomic obstruction was seen in 3 patients. In all 3 it was a result of anomalous anterior insertion of the tension apparatus of the anterior (superior) leaflet of the left AV valve (Fig. 2).
Malalignment of the aorta was seen in 31 of the 49 patients (62%) in whom it could be calculated; the maximal value was 73% (Fig. 3). In 10% of the patients the malalignment ratio was less than 80%, indicating considerable rightward displacement of the aorta, often combined with an increased septal thickness. The distribution of the malalignment ratio is shown in Figure 4.
Passive muscular LVOT obstruction was found in 71% of the cases. The relative diameter of the LVOT was, at the onset of systole, 92% ± 27% for the whole group (percentage of the aortic root diameter) (range 35% to 143%). Ten percent of the patients showed an LVOT obstruction at the beginning of systole of up to one-third of the aortic root diameter. The distribution of this relative diameter is shown in Figure 5.

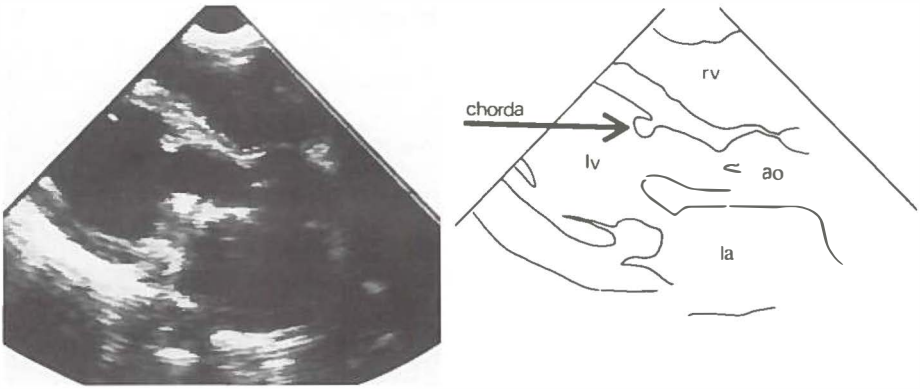

Fig. 2. Two-dimensional long-axis echocardiogram. The **arrow** points to an anomalous chorda floating in the left ventricular outflow tract. The mitral valve has been replaced by a mechanical prosthesis. ao = aorta; la = left atrium; lv = left ventricle; rv = right ventricle.

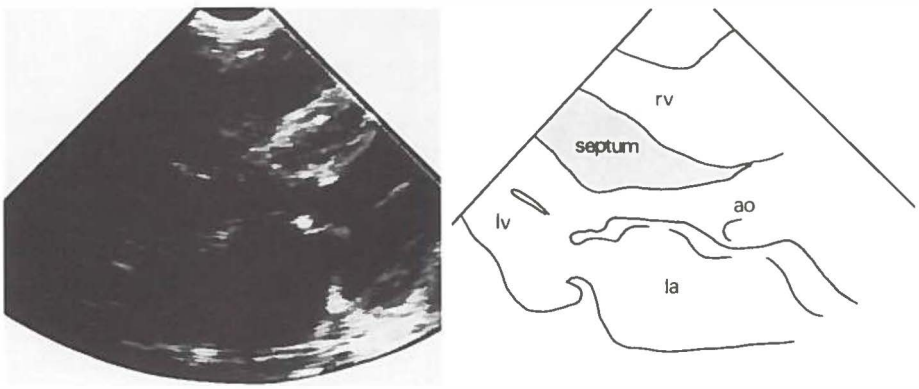

Fig. 3. Two-dimensional long-axis echocardiogram. The ventricular septum is shaded in the schematic drawing. The malalignment of the aorta (ao) over the septum is clearly visible. Note the markedly thickened septum. la = left atrium; lv = left ventricle; rv = right ventricle.

The muscular LVOT was further constricted in systole in all of the patients. The minimal relative diameter of the LVOT was 77% ± 22% (percentage of the aortic root diameter) (range 23% to 129%).
The sequential measurements showed a gradual decrease during systole to 83% ± 9% (range 67% to 100%) of the initial diameter of the LVOT. The velocity of narrowing showed 2 distinct periods of maximal velocity. These were in the first

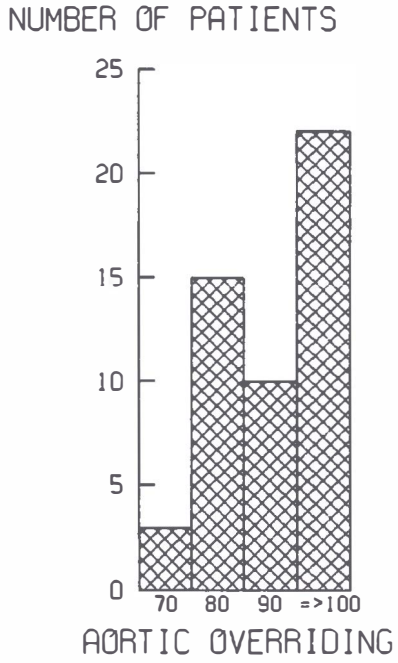

Fig. 4. Distribution of the aortic malalignment ratio. A small malalignment ratio indicates a considerable malalignment, whereas ratios of 100 or more indicate no malalignment.

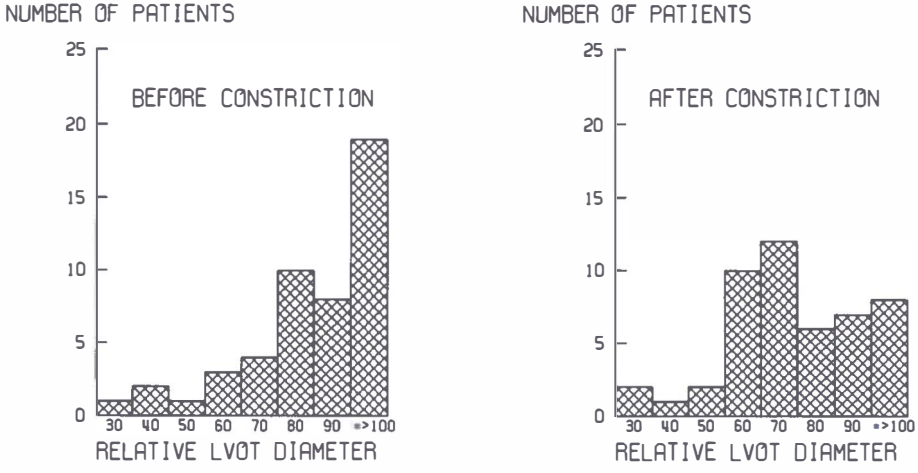

Fig. 5. Distribution of the relative diameter of the left ventricular outflow tract (LVOT). The diameter of the LVOT is expressed as a percentage of the diameter of the aortic root. The diameter clearly diminishes after constriction, i.e., at end-systole.

fifth of systole and towards its end (Fig. 6). The initial velocity of narrowing was much greater in patients who showed a minimal diameter of less than 83% ($p = 0.00001$) (Fig. 6).
Premature closure of the aortic valve was found in 16 of 58 patients (28%). These 16 patients had a significantly ($p < .05$) larger velocity of LVOT narrowing in the first fifth of systole than the other patients.

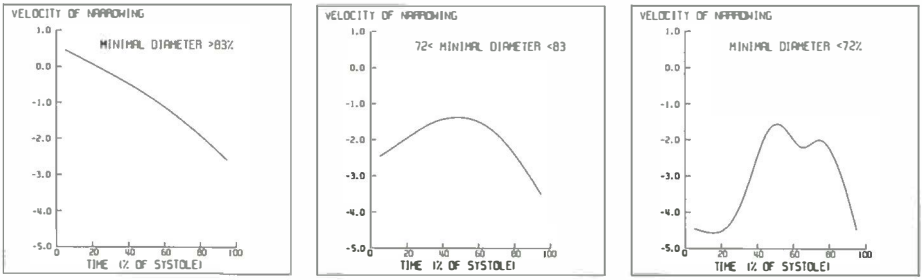

Fig. 6. Velocity of narrowing of the left ventricular outflow tract (LVOT) in 3 subgroups. The relative LVOT diameter is expressed as a percentage of the initial LVOT diameter. To determine the velocity of LVOT narrowing during systole, we calculated the first differential against time. The results of the calculation are shown for 3 subgroups. The subgroups differ in the minimal relative diameter of the LVOT. **Left,** the group with a minimal diameter 83%. **Middle,** the group with a minimal diameter between 72 and 83%. **Right,** the group with a minimal diameter of 72%. The graphs clearly show a difference in the velocity of narrowing in the first 20% of systole. Apparently, the initial velocity of narrowing is much greater in patients who show a minimal diameter of less than 83%.

Discussion.

It is well established that compared with the large subaortic outflow tract of the normal heart, that of an AV septal defect is intrinsically narrow. It is an elongated structure, made up posteriorly of an area of fibrous continuity between the aortic and AV valves and bounded on its other borders by the LV free wall, the ventriculo-infundibular fold and the outlet component of the muscular ventricular septum. There is no evidence to show that this intrinsically narrowed outflow tract produces organic stenosis during life. Indeed, symptomatic outflow tract obstruction is rare during life - much rarer than the overt lesions observed in autopsy studies that produce unequivocal narrowing.[3,4] Fixed obstruction was found in only 3 of our patients. Although our findings confirm the anatomic observations of intrinsic narrowing, the elongated and constricted outflow tract very rarely produces symptoms in patients who have undergone surgical correc-

tion of their AV septal defect; but our observations suggest that the LVOT, if capable of providing unhampered blood flow to the aorta, might do so with diminished reserve capacity. This is significant because Lappen et al[10] reported 5 patients in whom subaortic obstruction did not become manifest until after operation. Our findings point to the likely mechanisms that underlie this progressive stenosis and the means by which they may be predicted.

We have shown that, except in cases with fixed obstruction, the 2 major factors that narrow the LVOT are extreme rightward displacement of the aorta (malalignment) and "squeezing" of the tract between the septum and its fibrous posterior wall. We have also shown that there was constriction of the tubular outflow tract during ventricular systole.

This constriction shows 2 patterns. In most patients this muscular constriction was greatest at the very end of the systole, when most of the output has already been ejected into the aorta; but a second pattern was observed in which the outflow tract also narrowed significantly during the first fifth of systole. This second pattern was seen in patients with the greatest degree of aortic malalignment and with the greatest degree of squashing of the outflow tract between the intervalvar fibrous area and the septum. Additionally, this second pattern correlated with premature closure of the aortic valve, thus indicating unequivocal functional subaortic stenosis. These patients may be most at risk of organic stenosis.

The nature of the walls of the outflow tract and their pattern of contraction can readily be seen with echocardiography. The measurements in the present study were all made in patients after operation. As far as we know, the abnormalities described do not result in symptomatic disability, but the long term natural history of these patients is relatively uncharted.

Our results certainly show that they might be at risk. The resistance across the outflow tract relates to the fourth power of its diameter, assuming its cross section to be circular. Thus, anything narrowing the diameter by one- half will increase resistance 16-fold. Some of our patients seem much closer to a potential problem than others. We intend to investigate further the significance of our findings in these patients. Similar studies are also required in preoperative patients. Until these studies are completed, we can only state that a series of elements contribute to a potentially perilous arrangement of the LVOT in patients with AV septal defect. This intrinsically narrow tunnel was constricted during systole by its muscular walls, and this situation was exacerbated if the aorta was malaligned. Any degree of fixed stenosis could produce immediate overt obstruction. It may well be that the systolic murmur often attributed in these patients to AV valve regurgitation could also originate from the narrowed subaortic outflow tract.

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CHAPTER VI

THE SURGICAL ANATOMY OF THE LEFT VENTRICULAR OUTFLOW TRACT IN ATRIOVENTRICULAR SEPTAL DEFECT

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ABSTRACT

The left ventricular (LV) outflow tract (OT) in atrioventricular (AV) septal defect is an important structure that paradoxically is hardly ever seen by a surgeon. The LVOT is prone to develop obstruction following surgical procedures, such as left AV valve replacement, that seemingly do not affect the LVOT itself. We examined 15 hearts with AV septal defects and noted the anatomical boundaries of the LVOT. Additionally, the LVOT was examined microscopically, and it was sectioned to replicate echocardiographic images. A sham operation was performed to show the extent of the proposed resection for AV valve replacement. The mean length of this area was 91.8% ± 35.5% (range, 28.6% to 167.0%) of the diameter of the ascending aorta in our specimens of the Rastelli A variety. The mean diameter of the LVOT was 68.2% ± 13.5% (range, 42.9% to 100.0%) of the diameter of the ascending aorta. The posterior wall of the OT can either be resected or widened. Resection seems to be opportune at AV valve replacement, whereas widening could be performed when the OT is intrinsically stenotic. When one fully appreciates the concept of a five-leaflet common valve, it is clear that the length of the OT depends on the extent of adherence between the superior bridging leaflet and the septal crest. In hearts that have two separate AV valve orifices, the OT is fully developed; there is no potential for interventricular shunting ("ostium primum defect"), because the superior bridging leaflet is always tightly adherent to the septal crest. AV valve replacement in these cases is especially hazardous. Resection of the atrial fold (i.e., the posterior wall of the OT) might well be a solution to this problem.

Introduction

The left ventricular outflow tract in atrioventricular (AV) septal defect is a structure of considerable surgical importance. Paradoxically, its real anatomical proportions are hardly ever seen by the surgeon. It has more obvious anatomical boundaries in AV septal defect than in other congenital lesions. Indeed, its distinct morphology is considered to be pathognomonic for the lesion, usually being described as the "gooseneck" in a left ventricular angiogram[1]. Although the outflow tract itself is rarely sufficiently stenotic to produce clinical symptoms, it is known to be prone to rapid development of obstruction[2,3]. It is particularly well recognized that this treacherous occurrence can follow surgical procedures that seemingly do not affect the outflow tract itself, for example, replacement of the left AV valve. The high rate of mortality (25 to 40%) associated with this procedure could well be related to postoperative subaortic obstruction[4-6]. In other cases, outflow tract obstruction becomes manifest after closure of the interatrial communication[7].

As already stated, although the morphology of the outflow tract is readily delineated by echocardiography and angiocardiography, it is rarely seen by the surgeon using standard techniques. Both at primary repair and at AV valve replacement, the left atrium and the left AV valve are usually made visible in one of two ways: interatrial communication by way of a right atriotomy, or directly through a left atriotomy. The subaortic outflow tract cannot be seen with either approach. Furthermore, there is seldom a need to open the aorta. Therefore, the outflow tract remains literally in the dark to surgeons. The purpose of this study was to shed light on its surgical anatomy. We hoped to discover the anatomical basis of recognized surgical problems and propose techniques to relieve them or avoid them altogether.

Material and Methods

We examined 15 cadaver hearts with AV septal defect, taking particular note of the anatomical boundaries of the left ventricular outflow tract. The specifics concerning morphology of the bridging leaflets and their relation to the septum are shown in the Table. Histological sections at 0.5 mm intervals were made at right angles to the outflow tract in one heart that had separate AV valve orifices. These sections were stained with an Azan stain. Further hearts with separate and common valve orifices were sectioned to replicate the echocardiographic image as would be seen in the parasternal long axis view. A sham operation (described subsequently) was performed on one heart to show the extent of the proposed resection for AV valve replacement.

Results

Surgical Anatomy

To put the surgical anatomy in perspective in AV septal defect, we must start by comparing it with that of the normal heart. The normal left ventricular outflow tract is very short. Its posterior border is the area of fibrous continuity between the aortic and mitral valve leaflets. This area can be of variable length (and on occasion may be muscularized) but is usually just a few millimeters long. The outflow tract then achieves greater length in the areas of the commissures, which are well above the proximal attachments of the valve leaflets[7a,8]. Overall, however, the tract has little intrinsic length. In contrast, the left ventricular outflow tract in AV septal defect is much more extensive. An area of fibrous continuity (between the aortic valve and the left ventricular component of the superior bridging leaflet) exists that is much longer than the comparable area in the normal heart (Fig 1). Indeed, the mean length of this area was 92% ± 36% of the

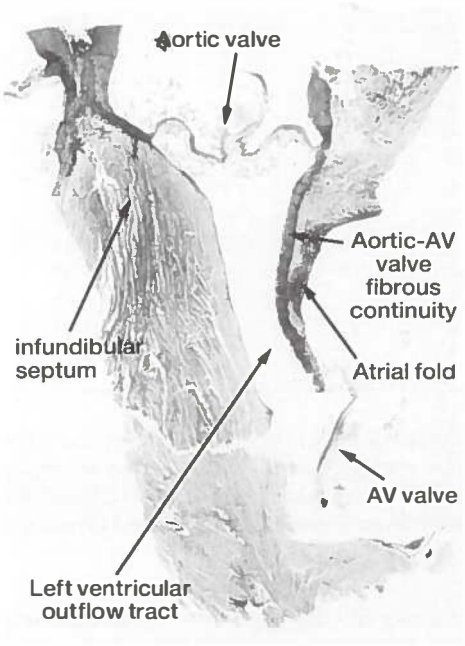

Fig. 1. Histological section of a left ventricular outflow tract in a plane similar to the echocardiographic parasternal long-axis view (Azan stain). The anterior and posterior limits of the outflow tract are visible. On the anterior side is the infundibular septum, and on the posterior side is the aortic-left atrioventricular (AV) valve continuity and the atrial fold. The "hinge" of the anterior bridging leaflet is clearly located more caudally than the aortic valve.

diameter of the aorta in our specimens of the Rastelli A variety (Table). Furthermore, its diameter was 68% ± 14% of the diameter of the aorta. On the atrial aspect of the area of the fibrous continuity we observed a muscular wedge. This is an integral part of the anterior wall of the left atrium and can be termed the atrial fold. It represents part of the inner heart curvature, being folded upon the area of intervalvar fibrous continuity. It is separated by these fibrous tissues and by the fibrous AV junction from the ventricular myocardial structures.

Table

Outflow Tract Measurements[a]

		Measurements (mm)			LVOT Calculated Values	
Patient No.	Rastelli Type	LVOT Length	LVOT Diameter	Aorta Diameter	Relative Length	Relative Diameter
1	A	10	8	11	0.91	0.73
2	A	6	5	7	0.86	0.71
3	A	10	6	8	1.25	0.75
4	A	6	3	7	0.86	0.43
5	A	5	4	7	0.71	0.57
6	A	4	3	5	0.80	0.60
7	A	4	3	4	1.00	0.75
8	A	10	5	8	1.25	0.63
9	A	5	4	7	0.71	0.57
10	C	1	6	8	0.13	0.75
11	C	1	9	9	0.11	1.00
12	A	10	4	6	1.67	0.67
13	A	4	8	8	0.50	1.00
14	A	9	6	8	1.13	0.75
15	A	2	5	7	0.29	0.71

[a]Measurements and calculations of the left ventricular outflow tract (LVOT) in atrioventricular (AV) septal defect. The relative length and the relative diameter are calculated as the proportion of the diameter of the ascending aorta. The patients with a Rastelli C type clearly have a shorter outflow tract. The relative diameter of the outflow tract is smaller than 1 in most patients.

In addition to the posterior fibrous segment, the anatomical boundaries of the left ventricular outflow tract are the ventricular septum and the left ventricular free wall (Fig. 2). When the superior bridging leaflet in AV septal defects is tightly connected to the septal crest, no structure is present that is comparable to the normal membranous septum. When a common orifice is present, the area of separation between the septal crest and the superior bridging leaflet may show a

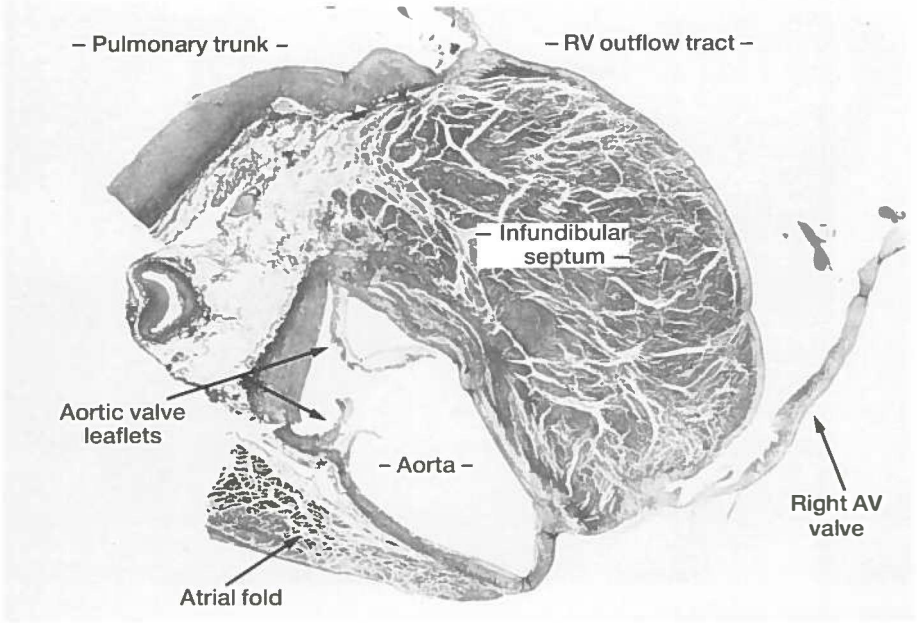

Fig. 2. Histological section of a left ventricular outflow tract in a plane similar to the echocardiographic parasternal short-axis view (Azan stain). The level is just on the caudal limits of the aortic valve. The extent of the outflow tract can be delineated. The atrial fold can be seen on the atrial side of the fibrous continuity between the left atrioventricular (AV) valve and the aortic valve. (RV ± right ventricular).

small triangular fibrous septum which is composed of the conjunction of a number of tendinous cords. The superior boundary of the tract in all hearts is the attachment of the aortic valve leaflets and, as in the normal heart, this is longer at the commissures. The inferior boundary is a discrete structure only posteriorly, being formed by the "hinge" of the superior bridging leaflet. In systole, this leaflet closes the AV orifice in a position at approximately a right angle to the outflow tract.

The preceding description is generally applicable to all AV septal defects. We noted, however, a marked difference in anatomy between hearts with separate right and left AV valve orifices and those with a common orifice (Fig 3). Because the attachment of the superior bridging leaflet to the septal crest is the inferomedial boundary of the outflow tract, the precise configuration clearly depends on the extent of this adherence. In all hearts with separate valve orifices, the superior bridging leaflet is tightly adherent to the crest of the ventricular septum as it crosses between the ventricles. In contrast, those hearts with a common orifice have superior bridging leaflets which are either completely free floating (as in

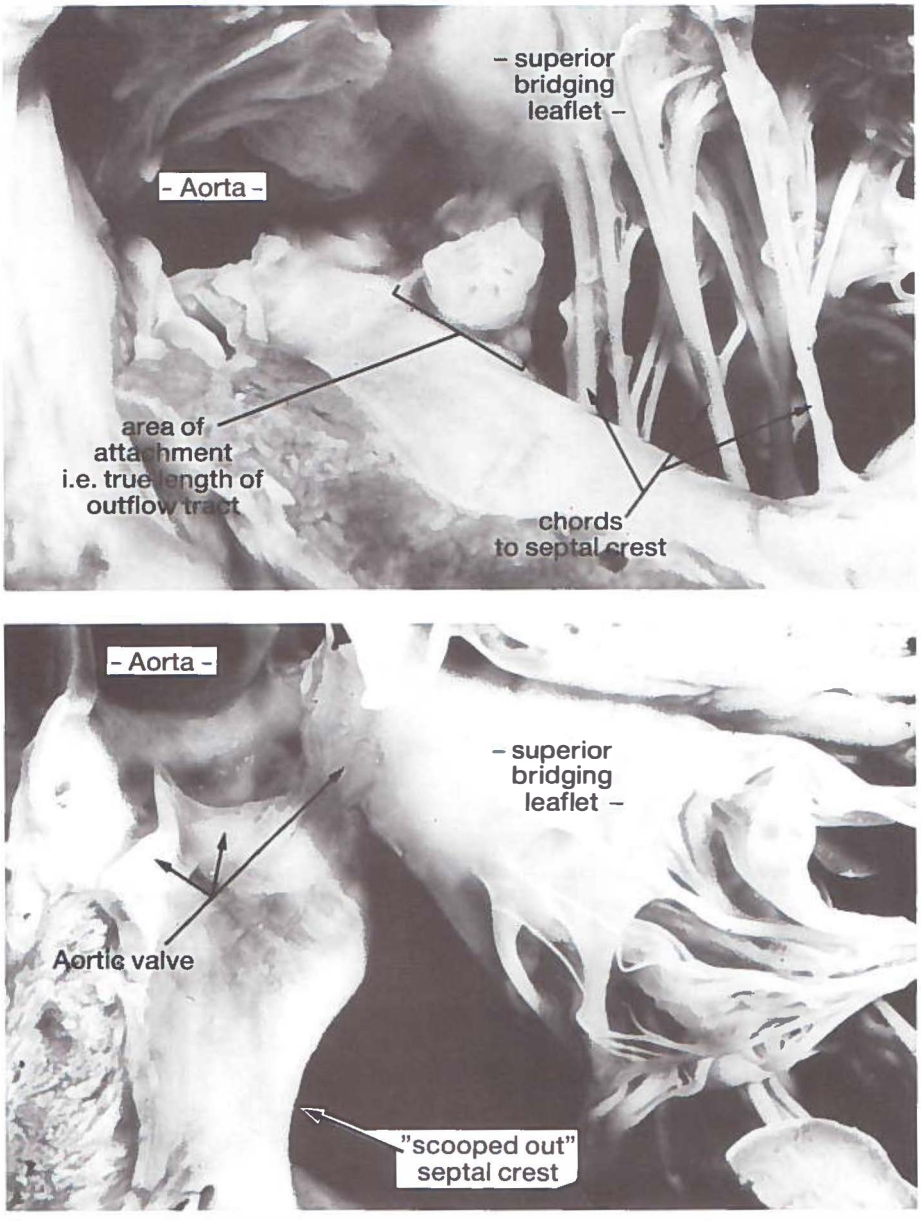

Fig. 3. Views of the left ventricular outflow tract from the lateral side. The difference in attachment between the superior bridging leaflet and the septal crest is shown between (A) a Rastelli A type and (B) a Rastelli C type. In the C type, the superior bridging leaflet is totally free floating. But in the A type tendinous cords attach the superior bridging leaflet to the septal crest. The bridging leaflet is partly directly connected to the septum, thereby creating the posteromedial boundary of the outflow tract.

the Rastelli C variety) or only partially adherent to the septal crest (as typically seen in the Rastelli A lesion). When the superior bridging leaflet is adherent to the ventricular septum, the inferomedial boundary of the outflow tract is elongated. But when the superior bridging leaflet is free floating, the outflow tract is considerably shorter. Then, in systole, the leaflet is "pushed" away from the outflow area toward the atrium. The outflow tract is therefore much longer when the superior bridging leaflet is tightly tethered to the crest of the ventricular septum (Table).

Surgical Implications

The atrial fold (the wedge of atrial muscular tissue) covers the extensive segment of valvar fibrous continuity on its atrial aspect. This contributes to a potential surgical problem. A surgeon looking from the atrial side sees a normal atrial wall. In our experience, the atrial fold has always been left in place when the AV valve has been resected for replacement by a prosthesis. The valve leaflet is usually removed at its "hinge" from the fold. When the AV valve is then replaced by the prosthesis, it is usually attached to the atrial fold. The fold then hangs like a curtain of tissue under the aortic valve. Together the prosthesis and fold

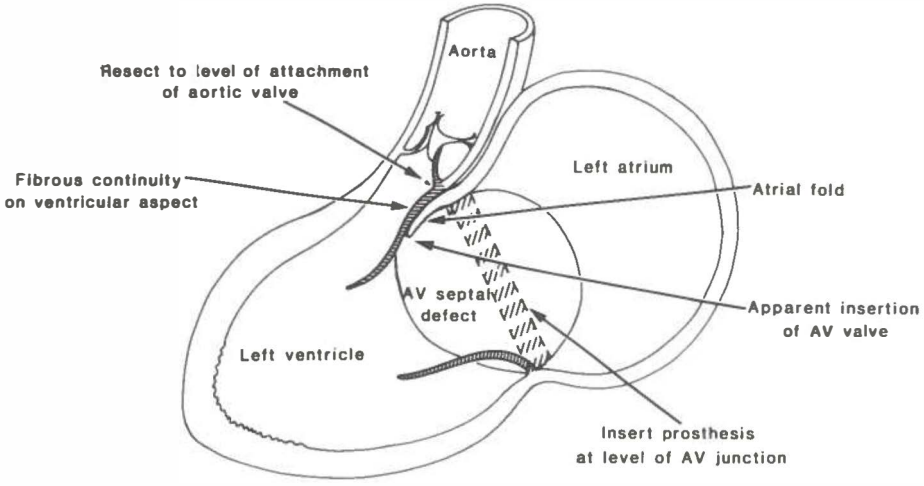

Fig. 4. Drawing illustrating the potential hazard of the insertion of an atrioventricular (AV) valve prosthesis. Usually the prosthesis is attached at the level of the apparent insertion of the AV valve. This causes a potential for outflow obstruction when, during systole, the prosthesis can be projected into the outflow tract. The prosthesis is drawn at the proposed level of insertion; the atrial fold should then be resected.

constitute a potential obstruction, particularly during systole, when they are likely to be pushed into the outflow tract (Fig. 4). This risk can be eliminated by resecting the atrial fold (Fig. 5). The prosthesis can then be attached at or close to the level of the aortic valve. Care should be taken not to extend the resection too far cranially because the aortic valve might be damaged or the heart might be opened into the transverse sinus of the pericardium.

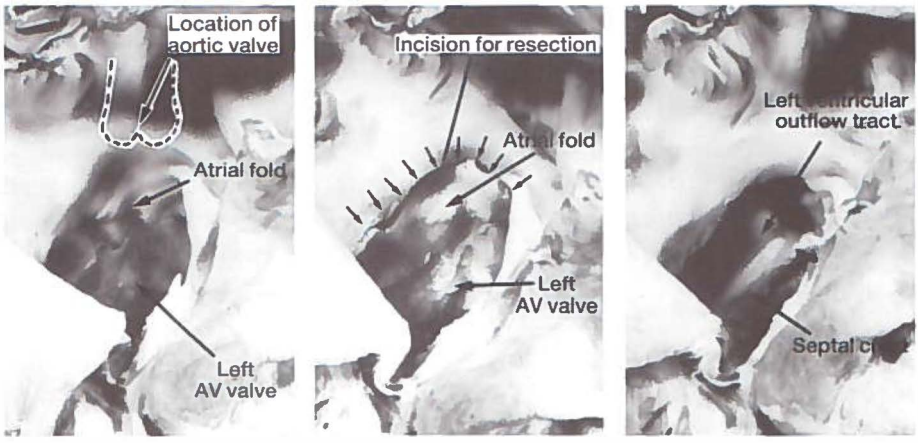

Fig. 5. A sham operation has been performed to show the proposed resection of the atrial fold at atrioventricular (AV) valve replacement. The photographs give a surgical view from the left atrium of the atrial fold, behind which the aortic valve can be found. In this example the valve and the atrial fold have been resected en bloc; in reality it might be easier to resect them separately. If the resection is extended too far cranially, one risks entering the transverse sinus of the pericardium or damaging the aortic valve.

Another operative problem relates to the means of providing relief of outflow tract obstruction when there is no need to replace the left AV valve. Lappen and co-workers[7] recommended and performed a septal myectomy in 3 patients and subsequent insertion of a pericardial patch in 1 patient to enlarge the outflow tract. The anatomy is such, however, that patch enlargement of the outflow tract is possible. This can be achieved by making an incision in the posterior wall of the outflow tract and extending it into the superior bridging leaflet. Insertion of a diamond-shaped patch will then enlarge the outflow tract (Fig. 6).

In hearts in which the superior bridging leaflet is totally free floating, the outflow tract is considerably shorter. As in the normal heart, the outflow length is limited primarily by the semilunar attachments of the aortic valve. After surgical repair in these patients, however, securing the patch between the septal crest and the bridging leaflet will create a new posteromedial border of a much more elonga-

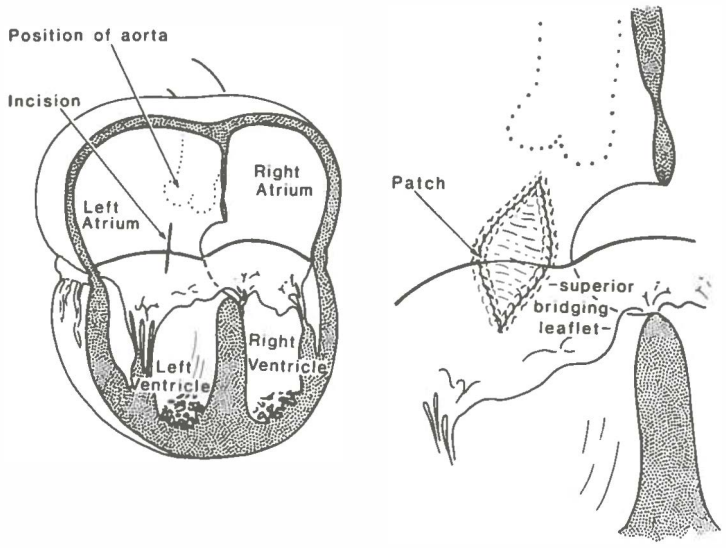

Fig. 6. Drawing showing the proposed patch enlargement of the left ventricular outflow tract from the atrial side. The diamond-shaped patch is extended into the superior bridging leaflet, analogous to a previously described technique for enlargement of the aortic annulus (Manouguian S, Seybold-Epting W: J Thorac Cardiovasc Surg 78:402, 1979[11]).

ted outflow tract. Depending on the position and size of the patch, this could then have the same potential for obstruction as occurs naturally in hearts that have separate valve orifices. It seems advisable, therefore, to attach this anterior part of the patch well on the right side of the septal crest, thus giving extra "width" to the newly created outflow tract (as is advisable on the posterior side to avoid damage to the conduction system).

Discussion

It is probable that neither the true nature of the left ventricular outflow tract in AV septal defect, nor the implications of its existence, have been fully appreciated. Behind an apparently normal atrial muscular wall is often found a surprisingly long and narrow outflow tract. This knowledge is of more than just academic value; neglecting it may cause morbidity and mortality[9]. An appreciation of the morphology of this outflow tract is difficult to achieve until one fully understands the concept of the five- leaflet common valve with superior and inferior bridging leaflets[10]. This arrangement occurs in hearts with a common valve orifice and in those with separate left and right valve orifices. Once this is known, it

is clear that the length of the outflow tract is variable; it depends on the extent of adherence between the superior bridging leaflet and the septal crest. The area of transition between the inlet and outlet septum curves cranially, and the superior bridging leaflet may or may not be attached to the superior component of this "scoop".

A heart that has two separate AV orifices and no potential for interventricular shunting "ostium primum defect" has an extensive connection between the septal crest and the bridging valve leaflets. The outflow tract is therefore always completely developed in these cases. However, hearts that have a common AV orifice have a variable anatomy. Usually the Rastelli A types have a more-or-less tight connection produced by tendinous cords extending between the superior bridging leaflet and the septal crest. The Rastelli C types have a totally free-floating leaflet. Intermediate morphologies of every degree are encountered, and indeed may rarely be found when there are separate right and left valve orifices. Generally, however, it is those with separate valve orifices (ostium primum defect) that have the most completely developed outflow tract. They are therefore in the greatest danger of outflow tract obstruction. But when a heart that has a common orifice is repaired, the danger then exists of creating an area of potential obstruction from a previously nonexistent or less well developed outflow tract. This is especially true if the superior bridging leaflet is stitched directly to the septal crest (rather than to its right ventricular aspect).

It could well be that the mortality often attributed to "low cardiac output" is associated with outflow tract obstruction. Preoperatively, the AV valve can be considerably regurgitant, in which case the regurgitant flow may increase the pulmonary flow and decrease the systemic flow (i.e., increase the shunt fraction). This happens even more readily in those hearts that have a common AV orifice when there is an extensive subvalvular (interventricular) communication. After repair, when the increment to pulmonary flow is removed, the systemic output usually increases. This may then convert a marginally obstructive outflow tract into an overt obstruction. Nevertheless, it is relatively uncommon for outflow tract obstruction to be manifest and documented after primary repair of AV septal defect. One reason for this may be that stenosis is neither suspected nor searched for.

A possible surgical solution, should stenosis of the native or newly created outflow tract be suspected, would be its enlargement from the left atrial aspect. This can theoretically be achieved by incising the bridging leaflet through the atrial fold. To avoid postoperative regurgitation, the patch should not be extended to the trailing edge of the superior bridging leaflet. Thus far, this procedure remains conceptual. It is similar to the patch enlargement of the aortic annulus into the aortic leaflet of the mitral valve as described by Manouguian and Seybold-Epting[11] Another surgical possibility is the one described by Lappen and associa-

tes[7] who detached the anterior bridging leaflet from its original attachment to the septal crest and inserted a patch into the gap.

Because of its high mortality rate (25% to 40%), AV valve replacement in AV septal defect is a problem of considerable magnitude. Reoperations for mitral valve replacement in the normally connected heart carry a considerably lower rate of mortality (8.7% reported by Bosch and colleagues[12]. An additional complicating factor clearly exists in AV septal defect. Obstruction of the subaortic outflow tract by the prosthesis has often been implicated. Crucial to understanding this complication is knowing the relationship between the aortic and atrioventricular valve. The "hinge" of the superior bridging leaflet is located much more caudally than is the attachment of the aortic valve. This is immediately apparent to morphologists, angiocardiographers, and echocardiographers, but it is not readily seen by surgeons.

If the valve is replaced by a prosthesis inserted at the "hinge" of the left ventricular component of the superior bridging leaflet, the prosthesis, especially if it is relatively large, will almost certainly project into the subaortic outflow tract. It makes more anatomical sense to suspend the valve much closer to the level of attachment of the aortic valve. The surgical problem is that the level of attachment of the aortic valve leaflets cannot be seen from the atrium. The true level becomes apparent only when the atrial fold (the posterior wall of the outflow tract) has been resected.

Resection of the fold and insertion of a valve at the approximate level of the aortic valve then eliminates the possibility of obstruction. To detect the approximate level of the aortic valve, one can probe from the atrium after temporarily releasing the aortic cross-clamp. Various suggestions have been made in regard to suturing the valve some distance above the AV junction within the left atrium. This procedure has been described by McGrath and colleagues[13], but it still leaves the atrial fold as a potential obstruction beneath the newly inserted prosthesis. This problem is avoided when the fold is resected.

In conclusion, the extent of attachment between the superior bridging leaflet and the septal crest is of crucial importance in determining the potential for obstruction of the left ventricular outflow tract in AV septal defect. It is this single factor which determines its length. In some circumstances the posterior wall of this extreme outflow tract can be resected. In others it could be enlarged by a patch. One should be aware of its hidden presence during any intracardiac surgical procedure in AV septal defect.

The authors thank C. Baarveld, D. Brekhoff, K. Kooistra and A. Winselaar for their technical assistance in preparing the histological sections.

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CHAPTER VII

LEFT ATRIOVENTRICULAR VALVE AFTER SURGICAL REPAIR IN ATRIOVENTRICULAR SEPTAL DEFECT WITH SEPARATE VALVE ORIFICES ("OSTIUM PRIMUM ATRIAL SEPTAL DEFECT"): AN ECHO-DOPPLER STUDY.

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Abstract

Left atrioventricular (AV) valve dysfunction is the most frequent major postoperative hemodynamic complication in patients with AV septal defect. The anatomy and function of the left AV valve were investigated in 64 patients with separate valve orifices (ostium primum atrial septal defect) who had survived corrective surgery. M-mode and cross-sectional echocardiograms of the left AV valve were obtained. Doppler flow tracings were obtained at the left AV valve orifice to determine if regurgitation was present. The findings were related to the position of the commissures between the leaflets, the size of the 3 leaflets and the position of the papillary muscles. Left AV valve regurgitation was present in 29 of 51 patients (57%). These patients had a significantly different left AV valve leaflet configuration, characterized by a large mural leaflet and a small inferior bridging leaflet. The size of the superior bridging leaflet is not a determinant factor. Thus, the configuration of the left AV valve in AV septal defect is related to the postoperative functional result. Awareness of the echocardiographic anatomy may influence the surgical approach to this defect.

Introduction

Left atrioventricular (AV) valve dysfunction is the most frequent postoperative hemodynamic complication in patients with AV septal defects.[1-3] Such dysfunc-

tion is promoted by 3 morphologic features. First, the left valve in AV septal defects has 3 leaflets, namely, the superior and inferior bridging leaflets and a mural leaflet.[4] Second, the papillary muscles are abnormally positioned. Third, valve function is compromised by an absolute deficit of leaflet tissue.[5] Because of this congenitally abnormal anatomy, the valve is prone to regurgitation. The regurgitation is presumed to take place particularly through the "cleft" between the bridging leaflets.

As Carpentier[6] pointed out, the surgical concept of suturing the so- called cleft in an attempt to create a mitral valve disregards the 3-leaflet nature of the left valve, which can never even resemble a normal mitral valve. In case of left AV valve dysfunction, instead of suturing together the bridging leaflets (which might immobilize the valve), he described an intricate reconstruction of the entire valve apparatus, including tailoring of leaflets, commissures and papillary muscles. Our study extends his concept by examining the relation between valve incompetence and the cross- sectional echocardiographic appearance of the valve leaflets and their papillary muscles after repair of AV septal defects with separate valve orifices ("ostium primum defects"). Our aim was to identify a specific morphologic feature that might be associated with a poor functional result.

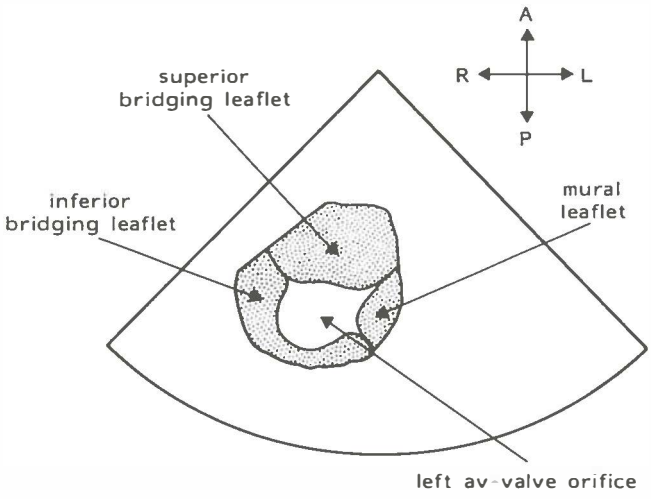
Methods

Patients. Seventy-five patients with AV septal defects with discrete right and left AV valve orifices underwent surgery between 1962 and 1982. The diagnosis was established by physical examination, chest radiography (increased cardiothoracic ratio, mean 59 ± 8%, range 47 to 76%), electrocardiography (increased PR interval, mean 178 ± 38 ms, range 100 to 260, and left-axis deviation, mean -60° ± 23°, range 0° to 120°) and cardiac catheterization with left ventricular cineangiography. In all patients, the atrial septal defect was repaired with an autologous pericardium-Teflon® double patch, and the cleft was closed to the extent that maximal competence of the valve was considered achieved. In many patients, abnormal tension apparatus attached within the outflow tract was removed. Sometimes mobilization of the inferior bridging leaflet was required because of abnormal chordal attachments.[7]

Sixty-nine of the 75 patients had survived until 1983 (92%) and 64 (85%) of these could be recalled for the echocardiographic study. For technical reasons, in 14 patients echocardiography did not supply all data required. The measurements that were obtained were used in the study.

All patients had complete evaluation of the left-sided AV valve, using the parasternal long- and short-axis views and an apical 4-chamber view.[8] We calculated the ratio of left atrial and ascending aortic dimensions from the M-mode study.

The position of the leaflets and their papillary muscles were recorded on videotape in the parasternal short-axis view. The position of the 3 leaflets and their junctions (Fig. 1) were marked on an acetate overlay and the papillary muscles were subsequently marked on the same overlay.



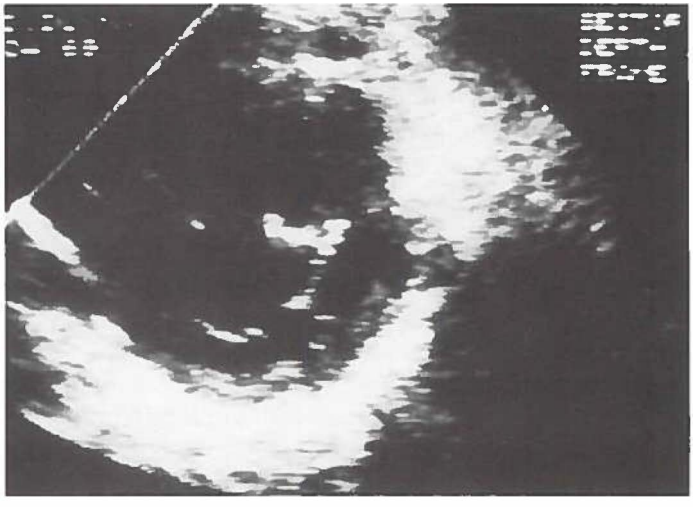


Fig. 1: A and B, cross-sectional short-axis echocardiogram of the left ventricle. Arrows point out the leaflets and the orifice of the left atrioventricular (av) valve. A = anterior; L = left; P = posterior; R = right.

We measured the relative sizes and positions of the left AV valve leaflets. The use of the interventricular septum as a reference point for our measurements was rejected, because its orientation and shape in AV septal defects are variable and assessment of its limits is highly subjective. Other fixed intracardiac points are not available in the short- axis view at this level. Because the left AV valve in AV septal defects is more circular in shape than normal hearts, we preferred to construct a midpoint by drawing 2 lines at right angles to each other. This divided the short-axis view into 4 equal quadrants. The only possible movement of the heart in the chest to interfere with these measurements is a rotation along the long axis. This possibility was evaluated and found not to exist.
It could be argued that choice of such a point could be influenced by the movement of the heart, but this is not the case for 2 reasons. First, the point itself is not floating because it is unique. Second, postoperatively the heart is fixed within the pericardium by connective tissue adhesion. The angles of the 3 commissures relative to anterior (0°) were measured from this calculated midpoint. From these measurements we calculated the angles of the sector that enclosed each leaflet. Finally, the angle between a line drawn through the 2 papillary muscles and the meridian of the drawing was determined. The distances measured from both papillary muscles to the midpoint of the interpapillary line defined the exact position of the papillary muscles on this line. The position of the interpapillary line itself was indicated by the distance from the midpoint to the centroid point on the meridian. There was no case of single papillary muscle.
With the transducer positioned to give an apical 4-chamber view, a Doppler flow tracing was obtained of the left-sided AV valve orifice. This tracing was used to document retrograde flow in systole indicating regurgitation through the left valve.
Additionally, to document the normal position of the papillary muscles of the mitral valve, we studied in similar fashion a group of 11 consecutive patients without heart disease.

Groups of patients were compared using a 2-tailed Student t test after checking for normal distribution. A p value < 0.05 was accepted as significant.

Results

Good-quality Doppler flow tracings were obtained in 51 of 64 patients. Regurgitation (visualized as jetlike negative velocity patterns, starting in early systole) was found in 29 patients (57%). No quantification or mapping was attempted. The group of 29 patients with Doppler-proved regurgitation had a larger left atrium-aortic ratio than those without ($p < 0.001$). The configuration of the left valve differed significantly between the groups with and without regurgitation.

Specifically, the sizes of the inferior bridging and mural leaflet differed significantly in the 2 groups. In contrast, there was no significant difference in the size of the superior bridging leaflet (Fig. 2). The difference in leaflet configuration, therefore, is caused by the position of the commissure and the concomitant sizes of the left ventricular components of the inferior bridging leaflet and the mural leaflet. The size of these leaflets is reciprocal. Thus, patients with regurgitation were characterized by a large mural leaflet, whereas those without regurgitation had a small mural and consequently a larger inferior bridging leaflet. No significant difference in prevalence of regurgitation was found between the group in which mobilization of the inferior bridging leaflet was required and the group in which it was not.

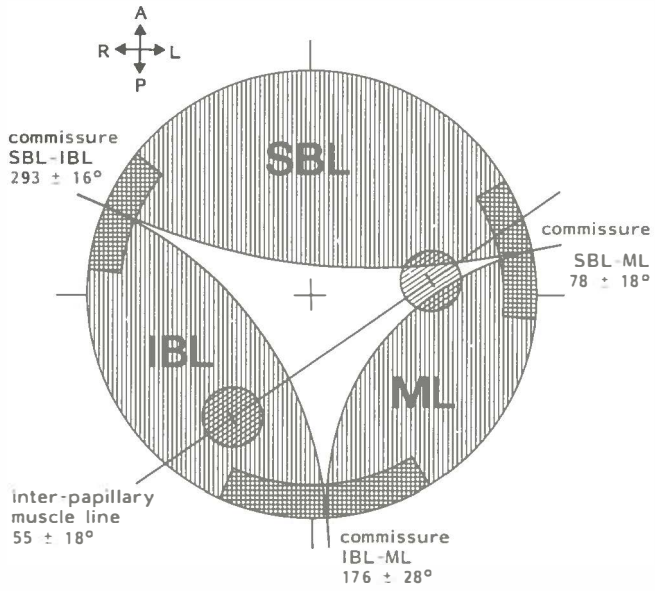


Fig. 2. The left atrioventricular valve orifice. Commissure and papillary muscles are depicted as mean positions, with their standard deviation. The largest variability is seen in the commissure between inferior bridging leaflet (IBL) and mural leaflet (ML). The posteromedial papillary muscle shows the most pronounced displacement. SBL = superior bridging leaflet; other abbreviations as in Figure 1.

The position of the papillary muscles differed significantly between the normal groups and the AV septal defects, counterclockwise rotation being found in the latter ($p < 0.05$). The posteromedial muscle was displaced away from the septum toward the lateral wall, whereas the anterolateral muscle was positioned normally (Fig. 3).

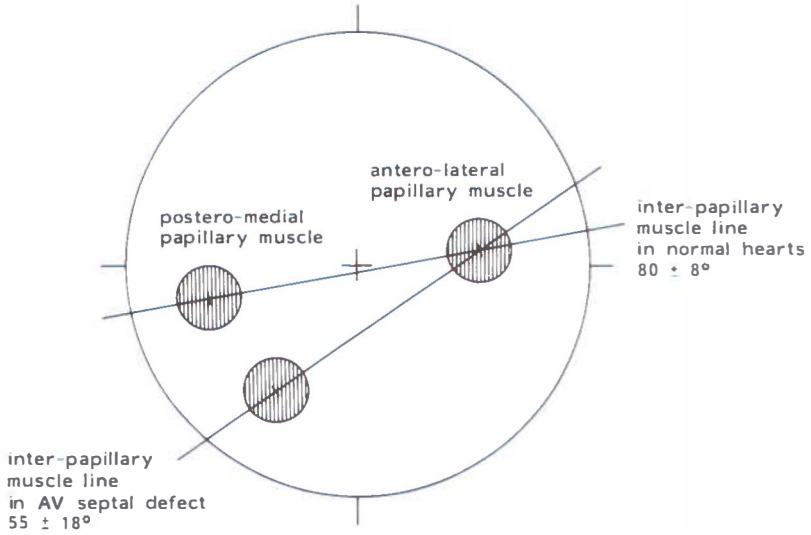


Fig. 3. The papillary muscle position of the left atrioventricular (AV) valve in the group studied and the normal group, depicted as mean positions ± standard deviation. The posteromedial muscle in the group with atrioventricular septal defects is displaced away from the septum toward the lateral wall.

Discussion

Our results suggest that in patients with AV septal defect, considerable differences exist within the setting of the trifoliate anatomy of the left-sided valve. The most variable feature was the position of the commissure between the inferior bridging and the mural leaflet, the other commissures being relatively constant in their position. The sizes of these leaflets were reciprocal, so that the inferior bridging leaflet was small when the mural leaflet was large and vice versa. The size of the superior leaflet was relatively unaffected by the relation between these other leaflets. We therefore presume that it is the size of the mural leaflet and the inferior bridging leaflet, respectively, that are crucial for proper function of the left-sided valve.

These anatomic findings are closely related to the function of the left valve. We assessed function by direct (Doppler-proved regurgitation) and indirect (left atrium-aortic ratio) methods. Both indicated that patients with a large mural and a small inferior bridging leaflet had regurgitation.

The left valve in AV septal defect resembles a mitral valve only in so far as it separates the left atrium from the left ventricle during ventricular systole. In all other respects, its architecture is profoundly different. A tongue of valve tissue

connects the 2 bridging leaflets when 2 separate AV orifices are present. In this way, in the presence of an ostium primum defect, a 3-leaflet left valve exists. The features observed in anatomic studies have been fully endorsed by our echocardiographic study. We observed a grossly different position of the papillary muscles, previously reported by Chin et al.[9] But, unlike Chin et al.[9], we found the posteromedial papillary muscle to be deviated. Our own finding is in keeping with anatomic observations.[5] We suggest that the difference in echocardiographic results relates to the mode of measurement used. The technique of Chin et al.[9] is different from our technique, because his "fixed" point of the septum in fact depends on the interpretation of the septal margins. Our centroid obtained by construction is unique to each heart, and thus our measurements more accurately reflect the different geometry between the normal heart and AV septal defects. The importance of the displacement of the posteromedial papillary muscle is just of anatomic interest. The variability in the angle of the interpapillary line showed no relation to the function of the valve.

A further question remains as to the use of the Doppler technique. In this study, we used this method solely in a qualitative role. Even in this role it provided crucial direct information over and above that provided by indirect means. Ideally, we would have preferred to have performed mapping and provided quantitative data, because there is good evidence that these are as reliable as angiographic data.[10] However, during this study, our efforts in this direction were insufficiently precise to provide accurate and reproducible results. The degree of regurgitation might correlate with the size of the mural leaflet. Nonetheless, our aim is to study this problem further in prospective fashion in patients both before and after operation. In a similar fashion, we have been unable to visualize and quantitate the amount of central leaflet tissue in the left valve during ventricular systole.[5]

The surgical significance of the anatomic features described have been elegantly discussed by Carpentier.[6] He devised a surgical technique for tailoring the left valve in such a manner that regurgitation is abolished (or at least minimized) while preserving its 3-leaflet anatomy. Unfortunately, even using this approach, perfect function in the left valve cannot be achieved in all patients.

The implication of our findings for prospective treatment therefore remains uncertain. Williams et al[11] recently described favorable results with individualized treatment in patients with AV septal defect with a common AV orifice, whereas Rizzoli et al[12] documented the improvement possible with adoption of the 3-leaflet concept for repair. We see no reason why our results in patients with separate valve orifices should not apply also to patients with a common orifice, since the basic leaflet configuration is the same.[4] How these findings might assist the surgical treatment of patients with an AV septal defect is still uncertain.

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LEAFLET SIZES AND COMMISSURE LOCATIONS OF THE LEFT ATRIOVENTRICULAR VALVE IN POSTOPERATIVE ATRIOVENTRICULAR SEPTAL DEFECT WITH SEPARATE ORIFICES, WITH AND WITHOUT DOPPLER DETERMINED REGURGITATION.

	REGURGITATION		STUDENT	P
LEAFLET	WITHOUT N=22	WITH N=29	T	VALUE
SUPERIOR	147° ± 19°	144° ± 17°	0.63	N.S.
MURAL	85° ± 28°	105° ± 26°	2.63	0.011
INFERIOR	127° ± 21°	110° ± 28°	2.38	0.021
COMMISSURE LOCATION				
SUPERO-INFERIOR	294° ± 15°	293° ± 18°	0.18	N.S.
INFERO-MURAL	166° ± 24°	182° ± 29°	2.10	0.041
SUPERO-MURAL	81° ± 17°	77° ± 19°	0.79	N.S.

CHAPTER VIII

ANOMALIES OF THE LEFT ATRIOVENTRICULAR VALVE AND RELATED VENTRICULAR SEPTAL MORPHOLOGY IN ATRIOVENTRICULAR SEPTAL DEFECTS.

Tjark Ebels M.D.*, Robert H. Anderson B.Sc.,M.D.,F.R.C.Path.*, William A. Devine B.Sc.**, Diane E. Debich B.Sc.**, Patricia A. Penkoske M.D.*, and James R. Zuberbuhler M.D.* Division of Cardiology (*) and Pathology (**) Children's Hospital of Pittsburgh, Pittsburgh, PA, U.S.A. Short title: Morphometry in AVSD.

Abstract

One of the characteristic features of atrioventricular septal defects is a deficiency of the inlet part of the ventricular septum, resulting in a "scooped out" appearance. The depth of the scoop in relation to the disposition of the atrioventricular valves has been debated. So as to clarify the relation between the morphology of the ventricular septum and the disposition of the atrioventricular valves, we quantified these anatomical features in 151 autopsy hearts, seeking to ascertain if those features identified particular groups within the overall lesion. We found that 137 hearts had left atrioventricular valves with three leaflets. The left valve in the other 14 hearts exhibited a dual orifice, a 2-leaflet or 1-leaflet arrangement or else was imperforate. These anomalies could be analyzed in terms of a sequence of diminishing formation of the commissures.

It was also shown that 3-leaflet valves displayed a variability in which the angular size of the mural leaflet correlated negatively with that of the inferior leaflet. In some of the hearts with a common atrioventricular orifice, the bridging leaflets did not meet over the ventricular septum, thus creating a "gap". Its size corresponded to a deficiency of the combined inferior-mural leaflet complex. Hearts with an abnormal disposition of the left atrioventricular valve had their ventricular septum scooped to a greater extent than those with a common orifice, even though most of them had separate right and left atrioventricular orifices.

Introduction

Atrioventricular septal defects are recognizable as a separate group of congenitally malformed hearts because of absence of any normal atrioventricular septal structures[1]. This feature results in a characteristically abnormal morphometry of the entire ventricular mass[2-4]. The atrioventricular valvar complex is also abnormal, being spatially perpendicular to the septal structures. Regurgitation of the left atrioventricular valve has been shown to be one of the most important incremental risk factors for death subsequent to surgical correction[5], and recent evidence suggests that this regurgitation depends on the individual configuration of the valve[6]. Some evidence exists which relates the structure of the valve to the septal architecture. Thus, measurements of a large series of hearts have shown a deeper "scoop" within the ventricular septum when the atrioventricular junction is guarded by a common valve as opposed to separate right and left valves[3,7]. Whether these septal dimensions are also related to the configuration of the left atrioventricular valve is unknown. Furthermore, there is still no agreement as to how and if "intermediate" variants exist within this overall architectural pattern[8-10]. With all these considerations in mind we have extended our previous morphometric studies of ventricular septal and atrioventricular valvar morphology[4] to study the angular configuration of the leaflets of the left atrioventricular valve and their relationship, if any, to the septal architecture. We have also investigated any relationships of our morphometric findings to well-recognized malformations of this left valve[11,12]

Materials and methods

We studied all 151 examples of atrioventricular septal defect from the Cardiopathological Museum of Children's Hospital of Pittsburgh, including the 120 hearts studied previously by Penkoske and her colleagues[4]. The hearts were categorized as having separate right and left atrioventricular orifices or a common atrioventricular orifice according to the presence or absence of a fibrous connexion of leaflet tissue between the superior and inferior bridging leaflets. Special note was made of the leaflet pattern of the left atrioventricular valve. In addition to determining the number of leaflets, we also measured their angular contribution to the entire 360° circumference of the left valve. These anatomical measurements were achieved by using special obturators manufactured so as to permit accurate measurement of the valvar morphometry (Fig. 1). Measurements of the ventricular septum were taken from the apex to three landmarks on its left ventricular aspect, the crux of the heart, the deepest part of the "scoop" and the superior attachment of the leaflets of the aortic valve (Fig. 2).
The measurements of the inlet and scoop were then divided by the outlet measu-

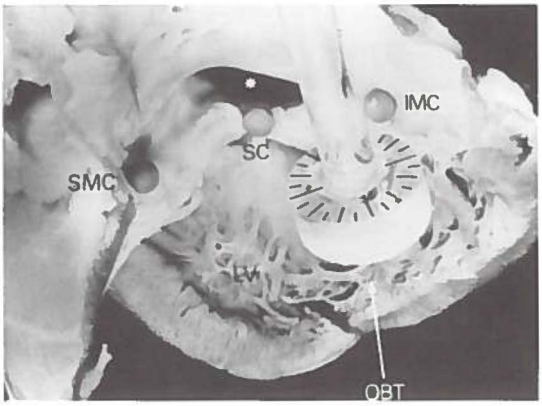

Fig. 1. Left sided view of a heart with an atrioventricular septal defect. Left atrium and ventricle have been opened by an incision through the mural leaflet of the atrioventricular valve. The three commissures have been marked by pins, and the obturator is positioned at the level of the orifice. The asterisk is located in the defect.
LV: left ventricle. OBT: obturator. IMC: infero-mural commissure. SC: septal commissure. SMC: supero-mural commissure.

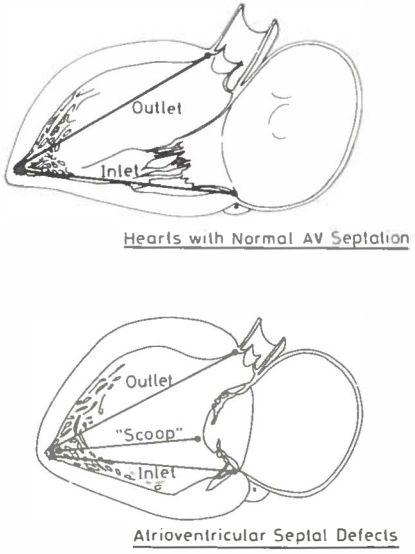

Fig. 2. Diagram showing the left side of the ventricular septum in a heart with normal septation and a heart with atrioventricular septal defect. The measurements taken from the hearts are shown in the bottom drawing: the outlet, scoop and inlet dimension. AV: atrioventricular.

rement and expressed as ratios, thus enabling direct comparison of one heart with another. Seventy percent confidence limits were calculated of binomial distributions. The significance of the difference between the means of measurements in different groups was tested using Student's T-test. The Chi-squared test was used for contingency tables. Analysis of variance was used for Table 3. A p-value of less than 0.05 was considered to be significant.

Results

On our first examination, we found 148 hearts in the museum having an atrioventricular septal defect. A common atrioventricular orifice was present in 103 hearts (70%; CL*: 65% - 74%) and separate left and right atrioventricular orifices in the other 45 hearts (30%; CL: 26% - 35%). Amongst these hearts, the left atrioventricular valve had three leaflets in 137 (93%; CL: 90% - 95%) (Fig. 3). The pattern differed from this usual 3-leaflet arrangement in the other 11 hearts (7%; CL: 5% - 10%). Three abnormal patterns were recognized (Table 1). A dual orifice was observed in seven hearts; a 2-leaflet valve in three; and a 1-lea-

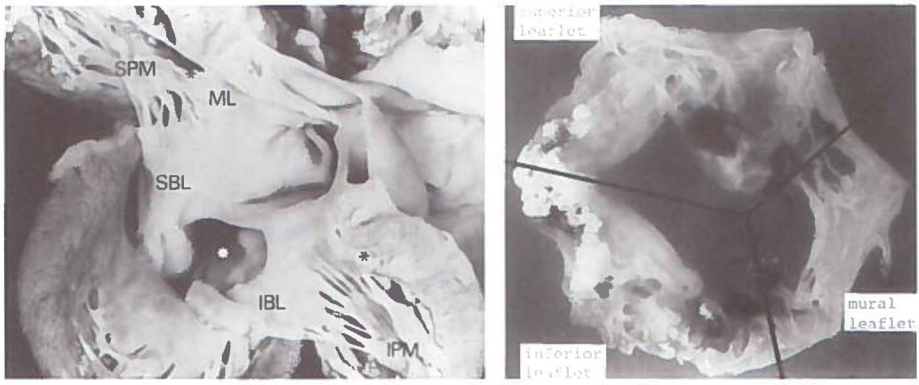

Fig. 3.

a. Left sided view of an atrioventricular septal defect, showing the trifoliate arrangement of the atrioventricular valve. *: defect. IBL: inferior bridging leaflet. SBL: superior bridging leaflet. ML: Mural leaflet. IPM: inferior papillary muscle. SPM: superior papillary muscle.

b. Ventricular view of a left atrioventricular valve from an atrioventricular septal defect, showing the trifoliate arrangement and the way in which the angular size of the leaflets was measured. Calcifications at the septal commissure are due to prior closure of the "cleft". The lines are drawn through the commissures and meet in the centre of the valve.

* CL: 70% Confidence Limits.

TABLE 1. TYPES OF AV VALVES IN AV SEPTAL DEFECT.

TYPE OF LEFT AV VALVE	NUMBER	% of 151	CL
3-LEAFLET	137	91%	88% - 93%
DUAL ORIFICE	7	5%	3% - 7%
2-LEAFLET	3	2%	1% - 4%
1-LEAFLET	1	1%	0% - 2%
IMPERFORATE MEMBRANE	3	2%	1% - 4%
TOTAL	151		

Legend: CL: 70% Confidence limits

flet arrangement in one other. The frequency of these less usual types of the left atrioventricular valve (being any other pattern than the usual 3-leaflet one) was four times as high in the setting of separate left and right orifices as when there was a common atrioventricular orifice (Chi-square test: p=0.0128) (Table 2). The dual orifice in all seven hearts existed because of an abnormal connexion between two leaflets of a basically 3-leaflet valve. In 6 (86%; CL:59% - 98%) of these, the abnormal connexion was found between the inferior and mural leaflets (Fig. 4a). Only 1 (14%; CL:2% - 41%) existed because of an abnormal connexion between the superior and the mural leaflets (Fig. 4b). Of these seven hearts with dual orifice, three had a common atrioventricular valve and four had separate right and left valves. The three specimens with left valves having two leaflets all showed failure of formation of the commissure between the mural and the inferior bridging leaflets (Fig. 4c). A common valve orifice was found in one of these three hearts. In the valve with an apparently single leaflet of the left atrioventricular valve in the presence of separate orifices, the left valve having a

TABLE 2. INCIDENCE OF UNUSUAL LEFT ATRIOVENTRICULAR VALVES

ATRIOVENTRICULAR JUNCTION	N	ABNORMAL	% of 148	CL
SEPARATE	45	7	16%	10% - 23%
COMMON	103	4	4%	2% - 7%
TOTAL	148	11	7%	5% - 10%

Legend: CL = 70% Confidence Limits.
Abnormal: All hearts with left atrioventricular valves without the normal 3-leaflet pattern.
Chi square test: p = 0.0128.

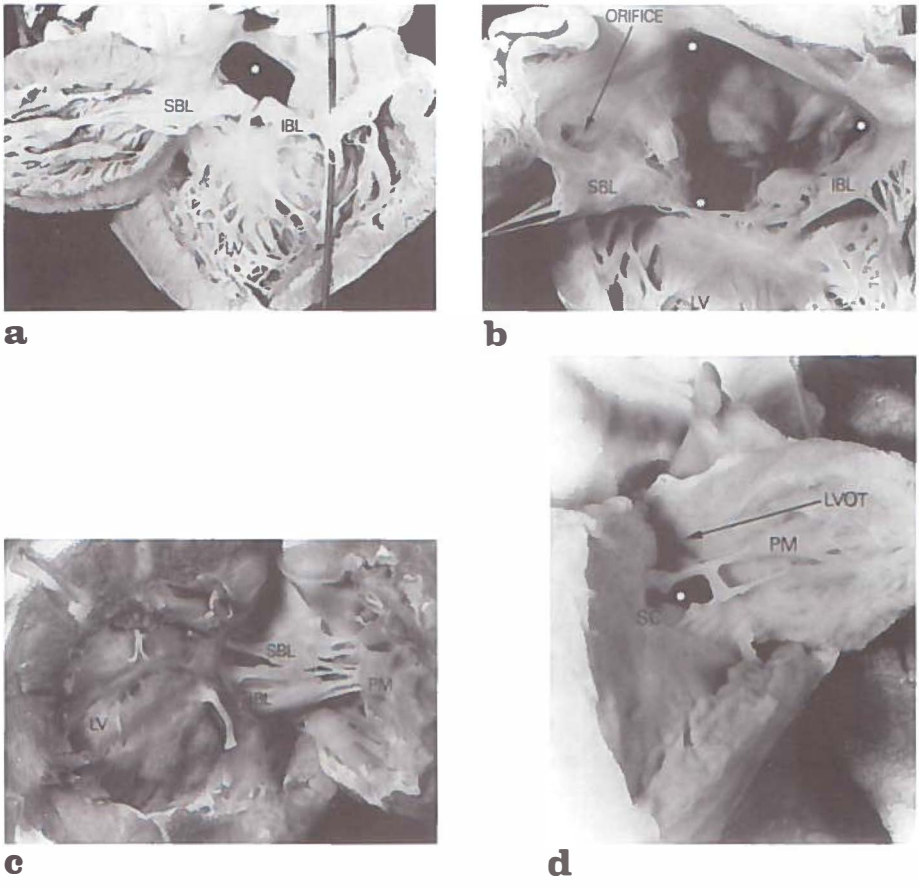

Fig. 4. Left sided views of atrioventricular septal defect with unusual atrioventricular valve morphologies.

a. An extra (double) orifice located at the site of the infero-mural commissure. A probe is sticking through the extra orifice. This is the usual location for an extra orifice.
b. An extra (double) orifice located at the site of the supero-mural commissure. This is a very unusual location for an extra orifice.
c. A bifoliate atrioventricular valve, supported by a single papillary muscle. There is no mural leaflet.
d. A single leaflet atrioventricular valve, having a (septal) commissure within itself. The valve is supported by a single papillary muscle and a solitary chorda.

*: defect. IBL: inferior bridging leaflet. LV: left ventricle. LVOT:left ventricular outflow tract. PM: papillary muscle. SBL:superior bridging leaflet. SC:septal commissure.

commissure within itself, there was no evidence of formation of discrete mural and bridging leaflets (Fig. 4d).
In all hearts with three leaflets, and also in those with dual orifice, the tensor apparatus of the left valve consisted of two papillary muscles which supported the juxtamural commissures. In those valves with extremely small mural leaflets, the discrete muscular heads supporting the commissural sites arose from a common body. The "extra orifice" in those hearts with dual orifice were supported by a smaller, but otherwise fully developed, papillary muscle. The valves with 2 leaflets or a solitary leaflet had a solitary papillary muscle within the left ventricle. In all those hearts with two muscles, they were positioned antero-superiorly and postero-inferiorly within the left ventricle rather than obliquely as is the case in normal hearts. The overall arrangement of the leaflets could be fitted neatly into a sequence of diminishing formation of the commissures leading eventually, it seemed, to an imperforate membrane (Fig. 5). Thus far, however, we had found no such imperforate variant. Further investigation in hearts within the museum

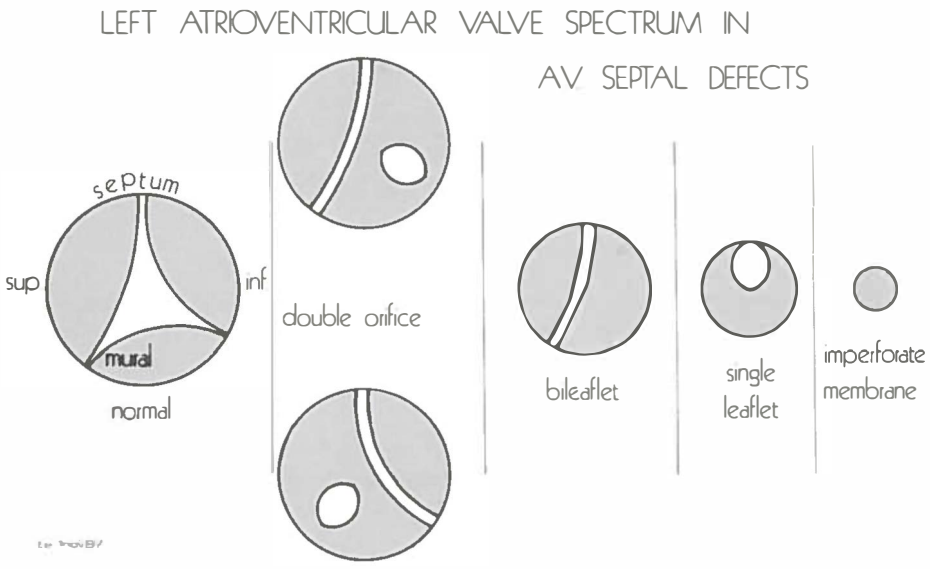

Fig. 5. Diagram of the sequence of atrioventricular valves in the setting of atrioventricular septal defect. On the left the "usual" trifoliate pattern is depicted, with superior, inferior and mural leaflet. Then, in a sequence of diminishing formation of the commissures, two types of double orifice valve, the extra orifice usually being located at the site of the infero- mural commissure (top). Unusually the double orifice is located at the site of the supero-mural commissure (bottom). Next the bifoliate valve, without a mural leaflet; a valve consisting of a single leaflet, with a commissure within itself. Finally at the right hand side of the spectrum the imperforate membrane.
Inf.: inferior. Sup.: superior.

then revealed three hearts having right atrial isomerism, atrioventricular septal defect, and diminutive left ventricles. These ventricles were found only after thorough dissection of the lateral wall of the ventricular mass. They proved to contain all the essential ventricular structures, albeit in rudimentary form. Within one such ventricle, with an "inlet length" of 6 mm., we found a single papillary muscle connected to an imperforate valvar membrane (Fig. 6). There was no outlet from this ventricle.

Fig. 6. Left sided view of the 6 mm left ventricle located within the left lateral wall of a heart with right atrial isomerism and atrioventricular septal defect. The imperforate membrane separating the ventricle from the atria is supported by a single papillary muscle.

In the overall group of hearts having three leaflets, the mean angular sizes of the leaflets were not significantly different (Fig. 7): mural leaflet 120° (SD**: 23°; range: 60° - 210°), superior leaflet 117° (SD: 16°; range: 90° - 150°), and inferior leaflet 113° (SD: 23°; range: 60°- 165°). The variance of the angular size was greatest between the mural and inferior leaflets, the variance of the angular size of the superior leaflet being significantly smaller ($F = 2.16$: $p < 0.001$ for difference in variances). No significant difference was found in the angular sizes of the leaflets between hearts having a common orifice or those with separate right and left orifices.

** SD: Standard Deviation

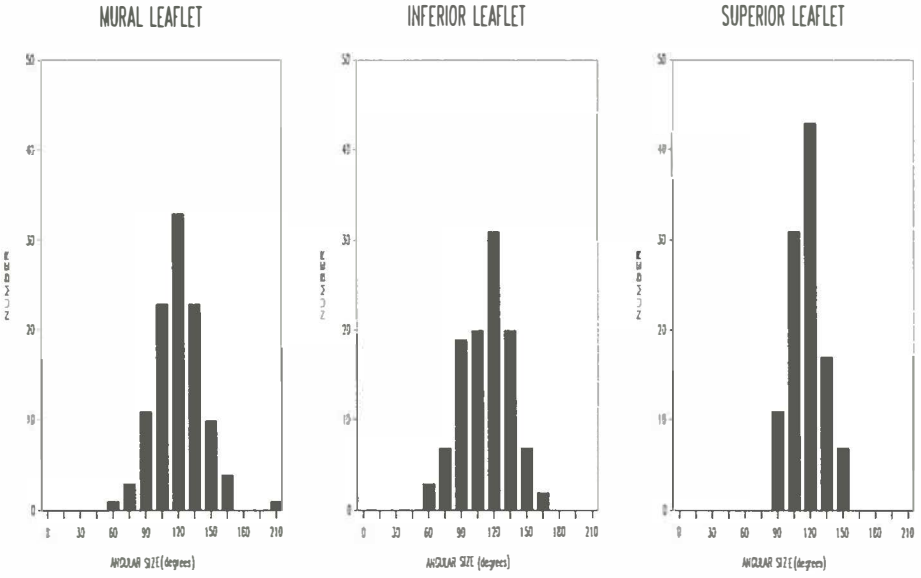

Fig. 7. Bar-graph of the distribution of the scoop/outlet ratio (horizontal axis) for the atrioventricular septal defects with a common atrioventricular orifice and those with separate left and right atrioventricular orifices. The hearts with common orifices clearly have a deeper scoop than those with separate orifices. There is considerable overlap, however, between the two groups.

The bridging leaflets coapted in 62 of the 103 hearts with a common atrioventricular orifice (60%; CL: 55% - 66%). In the other 41 (40%; CL: 34% - 45%), there was a gap between the bridging leaflets where the orifice could not be covered by leaflet tissue. The median size of the "gap" was 15°, but the maximum, measured in one case, was 75° (Table 3). Although the mean size of the

TABLE 3. THE ANGULAR SIZE OF THE "GAP" BETWEEN SUPERIOR AND INFERIOR BRIDGING LEAFLET IN HEARTS WITH ATRIOVENTRICULAR SEPTAL DEFECT AND COMMON ATRIOVENTRICULAR ORIFICE.

ANGULAR SIZE	NUMBER	%	CL
0°	62	60%	55% - 66%
15°	25	24%	20% - 29%
30°	7	7%	4% - 10%
45°	6	6%	3% - 9%
60°	2	2%	1% - 5%
75°	1	1%	0% - 3%
	------- +		
	103		

CL = 70% Confidence Limits

leaflets was similar, when the angular sizes of the leaflets were compared to each other in each case there was a very significant negative correlation between the angular size of the mural and inferior leaflet (r-0.60; $p<0.0001$) (Appendix). The angular size of the gap between the bridging leaflets had a highly significant negative correlation with the combined angular size of the mural and inferior leaflets ($r=-0.65$; $p<0.0001$) (Appendix).

Additionally, we found a poorly predictive, but very significant ($r= 0.37$; $p=0.0002$) positive correlation between the combined size of the mural and inferior leaflets and the ratio of the outlet and scooped septal dimensions. The size of the gap also showed a negative correlation with the scoop/outlet ratio ($r=-0.42$; $p=0.0001$). Comparison of the ratios of the inlet and outlet dimensions of the septum showed no difference between the groups having common and separate atrioventricular orifices. In contrast, when the mean ratio was calculated between the extent of septal scooping and the outlet dimension, a very significant difference was observed between the groups (T-test: $p<0.0001$), the scoop being greatest in the hearts having a common atrioventricular orifice.

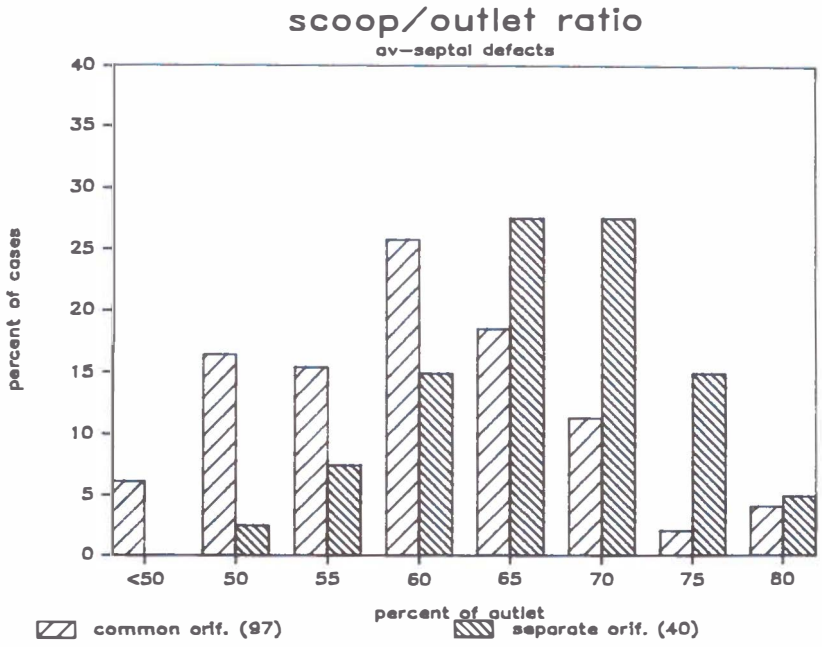

Figure 8.

Bar-graph of the distribution of the scoop/outlet ratio (horizontal axis) for the atrioventricular septal defects with a common atrioventricular ori- fice and those with separate left and right atrioventricular orifices. The hearts with common orifices clearly have a deeper scoop than those with separate orifices. There is considerable overlap, however, between the two groups.

Both groups displayed a normal distribution, with a considerable overlap between them (Fig. 8).
Of the hearts with unusual left atrioventricular valves, those with an imperforate or 1-leaflet valve had hypoplastic left ventricles, making measurement of the septal dimensions impossible. Only the 7 hearts with dual orifices and the 3 hearts with 2-leaflet valves possessed left ventricles sufficiently large to enable us to make accurate measurements. These 10 hearts with an abnormal left valve had a septal anatomy that was significantly abnormal compared to the overall group of atrioventricular septal defects. The scoop was deeper ($p=0.007$) and the inlet/outlet ratio was smaller ($p=0.008$) than in those hearts with a 3-leaflet valve, either in the setting of a common orifice or with separate orifices (Table 4).

TABLE 4. MEAN SEPTAL DIMENSIONS ON THE LEFT SIDE IN ATRIOVENTRICULAR SEPTAL DEFECT, EXPRESSED AS RATIO'S OF THE OUTLET DIMENSION.

ORIFICE TYPE	NUMBER	mean ± SD	
		INLET	SCOOP
SEPARATE	35	0.78 + 0.07	0.67 + 0.07
COMMON	97	0.76 + 0.07	0.60 + 0.08
ABNORMAL	10	0.68 + 0.11	0.56 + 0.11

Analysis of variance: Scoop/outlet ratio $p=0.008$
Inlet/outlet ratio $p=0.007$
Abnormal: Hearts with a dual orifice or 2-leaflet left atrioventricular valve.
SD: standard deviation

Discussion

It is now well-established that the ventricular mass in hearts with atrioventricular septal (canal) defects differs markedly from the normal[1-3]. In this study, we have extended our earlier observations concerning septal morphology and the basic configuration of the left atrioventricular valve[4] to concentrate on the precise morphometry of the valvar leaflets and their relationships to septal dimensions. We have identified a spectrum of malformations of the left valve extending from the typical arrangement with three leaflets, through a group of hearts with dual orifice, to those having two leaflets, one leaflet and, eventually an imperforate valvar membrane. Although abnormalities of the left valve are well-recogni-

zed[11-14], we are unaware of any previous account of this orderly progression of malformations, which can readily be interpreted in terms of disturbance of the anticipated 3-leaflet archetype. That the left atrioventricular valve in atrioventricular septal defects has three components is no longer in dispute. What remains contentious is the best way of describing these components. Our view is that each is best considered as a discrete leaflet, this contention endorsing the earlier opinions of Peacock[16] and Carpentier[17]. Our present observations strengthen our previously stated conviction that the valve is better not described as "mitral"[7]. In this study, we identified atrioventricular septal defects having left valves with only two leaflets and these bore no resemblance to the left atrioventricular valve as seen in the normal heart. As far as we can gather, the term "mitral" as applied to the left atrioventricular valve of the heart stems from Vesalius and was first used in 1543[18]. He likened the normal left atrioventricular valve to the top of an episcopal mitre. But, in 1880, the Austrian anatomist Joseph Hyrtl had criticized strongly this usage, even for the normal heart, stating that "the bishop's mitre came into the heart like the devil into the holy water font" (our translation)[19]. We can but echo this lament when "mitral" is used for description of the left valve in atrioventricular septal defect. Such semantic considerations, still likely to be highly contentious, should not detract from the potential surgical significance of these anatomic observations. When the left valve has three leaflets, there is considerable variability in the size of its mural and inferior leaflets. Our measurements show that, the larger the left ventricular component of the inferior bridging leaflet, the smaller is the extent of the mural leaflet and vice-versa, thus endorsing in an autopsy study earlier echocardiographic findings 61. Another study performed during life suggested that regurgitation was more likely to occur through the infero-mural commissure when there was a large mural leaflet[20]. Not all left atrioventricular valves were found to have three leaflets. Indeed, as discussed, a spectrum of valvar malformation was identified extending from the typical 3-leaflet pattern to hearts with an imperforate valve. Within this sequence, all seven examples we identified with dual orifices were produced because a tongue of leaflet tissue extended between the mural leaflet and one or other bridging leaflet. This had the effect of obliterating partially one of the commissures, the unobliterated component becoming the smaller of the two orifices. Such an interpretation of a valve with two orifices was first offered over 100 years ago by Greenfield in the setting of the normal mitral valve[21]. It would be dangerous to presume that, because the tongue is made up of normal leaflet tissue, it can safely be divided surgically. We would caution against such an approach. As far as we can judge, and as indicated by others[12], the valve would regurgitate massively should the tongue be divided. In general terms, nonetheless, the tensor apparatus in those valves with dual orifices is no different from that in the usual atrioventricular septal defect.

The papillary muscle which would normally support one of the juxtamural commissures now, in the setting of dual orifice, supports the "extra" orifice, albeit that in all our specimens the papillary muscle was smaller than its counterpart. It is worth emphasis that the papillary muscles in the overall group of atrioventricular septal defects occupy a fundamentally different location from those of the normal heart[4,6]. The left valves seen in those specimens with either two leaflets or a solitary leaflet confirmed the trend noted in commissural obliteration. Thus, in those valves with two leaflets, it seemed as if the commissure between the mural and one of the bridging leaflet had failed to develop. These valves, in keeping with this concept, were supported by a solitary papillary muscle (the so-called parachute arrangement)[12,13]. The valve with a solitary leaflet seemed to have failed to develop either mural commissure. It, too, was supported by a solitary papillary muscle (Fig. 4).

The orifice of this valve was, effectively, the septal commissure. The continuation of this spectrum was then seen in those hearts with an imperforate valve, the imperforateness seeming, to us, the consequence of failure of development of all commissures. All our hearts with abnormal left atrioventricular valves, therefore, could be interpreted in terms of alterations of formation of the 3-leaflet arrangement, this being the archetype of the left atrioventricular valve in atrioventricular septal defect. Our measurements have shown a significant but weak positive correlation in the overall group of hearts between the size of the combined mural and inferior leaflets and the degree of "scooping" of the ventricular septum. This, coupled with the relative constancy in the size of the superior bridging leaflet, lends some credence to recent evidence suggesting a different development origin of these components of the left valve[22]. In terms of the overall measurements, the small group of hearts with unusual left valves stood out from the overall group. They had a deeper scoop and a greater disproportion between their inlet and outlet dimension. These are the group which the Leiden investigators[23] have identified as a special group which they termed "intermediate". Although standing out as a discrete group, in no way are they intermediate between those having common orifices and those with separate right and left atrioventricular valves. On the contrary, they are more extreme in relation to both these patterns.

In view of the considerable confusion surrounding sub-classification of "intermediate" or "transitional" variants, our preference is to avoid these terms. Each group of investigators will have its own "shorthand" for describing these categories. Indeed it is our preference to avoid even the adjectives "partial" and "complete", describing instead atrioventricular septal defects either with separate atrioventricular orifices or a common orifice, and accounting separately for the anatomic potential for shunting of blood across the defect[4].

Appendix

Linear regression equations for relation between angular sizes of left atrioventricular valve leaflets and septal dimensions.

Mural leaflet° = 188° - 0.59920 * Inferior leaflet°

Correlation (R) = -0.60161; R^2=0.3619; T=-7.791; p<0.0001

Gap° = 129° - 0.51162 * (Mural leaflet + Inferior leaflet)°

Correlation (R) = -0.65213; R^2=0.42527; T=-8.559; p<0.0001

Scoop/outlet ratio = 0.29 + 0.00145 * (Mural leaflet + Inferior leaflet)°

Correlation (R) = 0.36636; R^2=0.13422; T=3.937; p=0.0002

Scoop/outlet ratio = 0.65 - 0.00213 * Gap°

Correlation (R) = -0.42338; R^2=0.17925; T=-4.650; p<0.0001

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CHAPTER IX

ECHOCARDIOGRAPHY AND SURGERY FOR ATRIOVENTRICULAR SEPTAL DEFECT

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Introduction

"Atrioventricular septal defect"[1] seems now to be well established as a suitably descriptive term for the spectrum of congenital anomalies that is the subject of two papers in the current issue[2,3]. "atrioventricular septal defect" unifies the spectrum of anomalies commonly called "ostium primum defect", "common atrioventricular canal", and "endocardial cushion defect". The obvious advantage of the purely descriptive "atrioventricular septal defect" is its lack of implications concerning embryological theories. These theories are subject to fashion and hence tend to change. The morphology does not. In describing morphology, therefore, we should not compromise ourselves with embryology, but stick to description. As an illustration, we probably cannot improve much on the first description of an atrioventricular septal defect by Thomas Bevill Peacock in 1846[4]:

> "The interesting points in the case were, first, a deficiency of the base of the inter-auricular septum, with a perfect closure of the foramen ovale. The deficient space allowed of free communication between the two auricles, so that there could only be said to be one auriculo-ventricular aperture. 2ndly. A distinctly tricuspid form of the left auriculoventricular valve; and 3rdly, a deficiency at the base of the septum of the ventricles, nearly closed by an extension of the anterior fold of the left auriculoventricular valve."

Two topics are of importance in relation to the unification of the concept of atrioventricular septal defect. First is the long standing realization that "ostium primum defect with a cleft mitral valve" and "complete atrioventricular canal" are

variants of one and the same anomaly. The only distinction is the well-demonstrated relation of the bridging leaflets of the atrioventricular valve with the septal crest[1]. The only valid reason to keep these two popular names was probably that their surgical treatment is different in complexity. Thus, many "ostium primum defects" can be treated as if they are "funny" atrial septal defects in which one has to suture a "cleft" in a mitral valve leaflet and be aware of the different arrangement of the conduction tissues. In contrast, an operation for a "complete atrioventricular canal" requires much more surgical dexterity and, as a consequence was, sadly, associated with an ominously high mortality. As a result of this sometimes prohibitive mortality, some paediatric cardiologists have argued that surgical treatment is not warranted in children with Down's syndrome, because they maintain (wrongly in my opinion) that life expectancy is not prolonged[5]. Nevertheless, it has been well established that the two anomalies are one in basic morphology, although they differ in certain surgically crucial (but anatomically minor) features[1].

The second topic, as a consequence of the first argument, is the name that we are to give to this group of anomalies. Two embryologic structures have been implicated in being anomalous in the development of this lesion, the developing atrioventricular junction ("canal"), and the endocardial cushions. The discouraging facts are that the atrioventricular junction in the young embryo bears scant resemblance to the definitive lesion and that the endocardial cushions make up very little of the formed heart. It now appears that, during normal development, the endocardial cushions rather glue together two parts of the aortic (anterior) leaflet of the mitral valve. And it might well be, therefore, that the only true endocardial cushion defect is the isolated cleft in the anterior leaflet of the mitral valve, a lesion which is unrelated to atrioventricular septal defect[6]. If so, this signals the defeat of embryologically based morphological names. But a single common denominator in this entire group of lesions is the absence of any atrioventricular septal structures, albeit that this is not the only abnormality as the heart morphology at large is distinctly abnormal. Thus, "atrioventricular septal defect" seems to be the best name, although one should realize that several suffixes are then needed to fully describe a specific case.

One of the other features common to all atrioventricular septal defects is the basic morphology of the atrioventricular valve. The entire valve apparatus is composed of five leaflets, of which three are committed in whole or part to the left ventricle. The superior and inferior leaflets bridge the ventricular septal crest, the former to a variable degree. One can distinguish a common orifice or two separate ones, the latter existing when the two bridging leaflets are connected by a "tongue" of leaflet tissue. Far more important to the surgeon is the connexion between the bridging leaflets and the septal crest. When there is a tight and complete connexion of both leaflets to the septal crest, there is no potential for inter-

ventricular shunting ("ostium primum defect"). Repair can then be limited to the easily visible atrial side of the atrioventricular junction. But when the connexion between bridging leaflets and the septal crest is incomplete or nonexistent, the interventricular communication must also be closed.

The interventricular communication

Echocardiography should ideally be able to demonstrate or exclude with accuracy the existence of an interventricular shunt. Silverman et al.[2] have clearly shown this to be possible in their superbly illustrated paper. While observing the heart from the apical four chamber view, tilting of the transducer from a caudal to a cranial direction enables the entire extent of the defect to be assessed. The disadvantage of this procedure is that there is not one single cross-sectional image that shows the entire defect, but each frame shows either the superior or the inferior bridging leaflet. Therefore several pictures are needed to be sure that one has visualized the entire defect (or the entire sweep would need to be viewed on a display unit). Especially when the interventricular part of the defect is small, one might miss it. It is unfortunate that Silverman and his colleagues[2] give no figures on specificity and selectivity, but that might be beyond the scope of this particular paper. Apparently there is no echocardiographic window through which one can shoot parallel to the septum and thus hit both bridging leaflets and view the entire relation of the atrioventricular valve apparatus to the septal crest in one single frame. On the other hand I wonder whether it would not be more reliable to visualize the shunt flow with colour coded doppler. For instance, in a subxyphoid short axis view one should be able to see the jet of interventricular shunt blood coming towards the observer. In this way it might be possible to detect even small interventricular defects.
But then, Kirklin et al.[3] state that their operative mortality for all atrioventricular septal defects is now the same, irrespective of the presence of an interventricular communication. For "partial" defect their mortality is 0% (CL:0%-11)* and for "complete" defect it is 2% (CL:0.3%-8%). One can only applaud such superb surgical management, while realizing that very few surgeons have the extensive experience of Kirklin and his colleagues. Especially in a generalized training situation such as ours these results might be difficult to match. On the other hand, I am sure that these mortality figures reflect also meticulous preoperative diagnosis, in which echocardiography can play a decisive role. And, because I presume that few surgeons can match these mortality figures[3] in the presence of

* CL = 70% confidence limits

an interventricular communication, I feel that the demonstration or exclusion of such a communication is crucial to the surgeon. This is the more so when it is well known that the connexion of the bridging leaflets to the septal crest might consist of numerous small chords, leaving small spaces in between and thus hiding the extent of the defect. Because of this, I am not completely satisfied with the procedure described by Silverman[2], because, from still frames, one can not be entirely sure that one has not overlooked a small interchordal communication. I challenge the echocardiographers, therefore, to produce one view, which shows us the entire defect and the relation of the bridging leaflets to the septal crest in a single frame. It seems to be possible by gated magnetic resonance imaging[7], but this procedure might remain cumbersome for routine appraisal of an atrioventricular septal defect.

Atrioventricular valve anatomy

To establish the diagnosis of "atrioventricular septal defect" it is helpful to visualize the typical configuration of the atrioventricular valve. That the "cleft" is a commissure (albeit different from other commissures) because it is the meeting point of two valve leaflets is now widely accepted[8]. Even more important is the appreciation of the trifoliate nature of the left atrioventricular valve. This would be no more than a curiosity were it not for the fact that the left atrioventricular valve is often considerably regurgitant. In the past attempts were usually made to repair the valve by constructing a proper "mitral valve", closing the entire "cleft in the anterior leaflet" regardless of the individual anatomy. Surprisingly, this procedure resulted in a valve which functioned reasonably well in the majority of patients. Unfortunately perhaps, these results strengthened the belief that a mitral valve (that is a bileaflet structure like a bishops hood) could be made out of the trifoliate entity found in atrioventricular septal defect. The credit goes to Carpentier for pointing out that the valve is, and should stay trifoliate[9]. Repair should be based on this trifoliate concept. This fact is in no way refuted by the regurgitation occurring most often at the distinctly abnormal commissure between superior and inferior bridging leaflets, whence this commissure should probably be sutured. In many other cases, however, this is not the sole solution to abolition of regurgitation, since the leak can occur at one of the two other commissures, or in the center of the valve because of an absolute deficit in valve tissue. Indeed, in certain configurations of the valve leaflets, dogmatic suturing of the "cleft" makes the valve stenotic.
We did an echo-doppler study on patients that had been operated upon for their "ostium primum defect" in the period that the accepted technique was to suture the "cleft" to the extent that the valve was judged to be optimally competent[10].

We showed that, in the patients with a regurgitant left atrioventricular valve, the mural leaflet was significantly larger than in the group without regurgitation. One of our conclusions is that in the patients with a relatively large mural leaflet, suturing of the "cleft" is apparently not the procedure of choice. It might very well be that one of the other commissures needs attention in this group of patients. Indeed, we now have now some evidence that in patients with a large mural leaflet the site of leaking might be at the commissure between the inferior bridging and the mural leaflet. When the mural leaflet is large, though, the inferior bridging leaflet is reciprocally small. The commissure between those two leaflets is then located relatively close to the conduction system, thus adding further danger to the surgical approach.

Silverman's figure 10 marvelously shows us the trifoliate left atrioventricular valve[2]. The exciting thing about this precise visualization of the valve leaflets is that their relative contribution to the left atrioventricular valve might give us an indication of the way it should be surgically handled. This is vital preoperative information, because it is rather difficult to precisely measure the relative size of the valve leaflets during the operation in a cardioplegic situation**. It is clear from the results of Kirklin et al.[3] that left atrioventricular valve dysfunction is paramount for the morbidity and mortality of the lesion, since the severity of preoperative atrioventricular valve incompetence is an incremental risk factor that remains constant. This is the more so because late left atrioventricular valve replacement still carries a high reported mortality of 25%-40%[11,12]. Possibly this mortality is related to left ventricular outflow tract obstruction by the prosthesis. This can be avoided by resection of the posterior wall of the outflow tract[13].

The superior leaflet can bridge the septal crest to a varying degree, this variability being the basis of the Rastelli classification[14]. This classification has surgical technical importance but does not appear to be a discriminative risk factor in Kirklin's analysis[3]. Neither was it in the analyses of Studer and Rizzoli and their colleagues[15,16]. Nonetheless, one of the merits of this classification might be that it tells us something about the left ventricular outflow tract. When we did a study on the surgical anatomy of the outflow tract in atrioventricular septal defect we noted it to be long and narrow in the type A of Rastelli et al[13,14]. In contrast, it was short and wide in the type C of Rastelli et al. But when surgically correcting a type C of Rastelli one should realize that the antero-superior part of the subvalvar patch forms the medio-posterior wall of the outflow tract. The interventricular patch can thus cause outflow tract obstruction.

** One method to calculate the relative angle occupied by the mural leaflet is to measure the diameter of the valve orifice and the geometrical chord between the two mural commissures. Then: sin(angle/2) = chord/diameter.

The case of echo vs. catheterization

If echocardiography can so perfectly portray the anatomy of the lesion, we may wonder whether there is still a need for catheterization. While the study of Silverman et al.[2] is primarily a comparison between morphology and echo images, it shows clearly that echo is capable of convincingly clear cross-sections demonstrating the anatomy. What is now needed is a study on the specificity, selectivity and predictive value of echo in this and other lesions. In such a study, not only should the diagnosis be an independent variable, but also the surgically relevant anatomical features.
Additionally, inter- and intra observer variability should be assessed in order to make such a convincingly reliable case that the surgeon will not encounter any disagreeable surprises on the operating table. When these questions have been resolved satisfactorily, one might then argue that catheterization is not necessary in routine cases.
What then would be the remaining value of catheterization? The information we cannot get by echo are hemodynamic parameters such as the all important pulmonary vascular resistance. When this resistance is raised, it is well known that the operative risk increases. But an elevated pulmonary vascular resistance as a consequence of a large left-to-right shunt usually develops after some time. And, when we operate on all these children early as rightly advocated by Kirklin et al.[3], we hopefully shall always be ahead of this foreboding development. So, correcting all these children early would be another argument against catheterization.
Nonetheless, some children already have an elevated pulmonary vascular resistance very early in life. Some develop it independently of an early successful operation. Maybe these two groups are one in reality and then the operation might not alter mortality in this group of patients. Thus, there are still many "maybe's" to be answered. Because of this, I think catheterization remains essential. Therefore, with the current echocardiographic possibilities, I think catheterization of patients with an atrioventricular septal defects will be less of a diagnostic and more a scientific tool.

Surgery and statistics

Kirklin's paper is an impressive example of the value of retrospective patient oriented analysis[3]. It gives profound insight into the operative and postoperative risks of patients with an atrioventricular septal defect. I agree wholeheartedly with the conclusion that these patients should be operated upon early in life, preferably in their first year. That the severity of preoperative atrioventricular valve

incompetence is still a constant risk factor signals that we should focus on a better understanding of the function of the left valve and its repair. This seems to be one of the few problems that still influence mortality figures for Kirklin and his colleagues[3].

The most striking feat of this paper though, is to show that their statistical method predicted future results from present day less than perfect results. The current paper shows the validity of these predictions. I am anxious to know what the predictions are for lesions that are still somewhat difficult to cure, such as the hypoplastic left heart syndrome. The rather philosophical problem then arises wether knowledge of these predictions alters the attitude of the surgeon. If this is so, knowledge of the predictions might become a factor in the risk analysis. For example, when one would be aware of the prediction that the mortality for a certain operative procedure is to be halved in, say, five years, he/she would be prone to take a more liberal stance regarding the indications for operation. That could influence future results and thus invalidate the prediction. I am not implying that we should not make these predictions. Maybe in clinical matters, however, we should deal with the present day mortality figures and not with predicted ones.

Natural history and down's syndrome

I agree fully with the discussion of Kirklin et al.[3] relative to the study of Bull and her colleagues[5]. Kirklin et al. argue that the data of Bull are not a valid representation of the natural history in patients with coexistent Down's syndrome. The starting point of Bull's analysis is not birth. Their patient population, therefore, is a selected one and is not representative of all patients with Down's syndrome and atrioventricular septal defect. Furthermore, referring patients physicians are undoubtedly aware of the stance of the paediatric cardiologists in the Brompton Hospital towards this problem, giving it another source of bias. Thus the conclusions of this paper are based on a favorable subgroup of the population that was probably referred specifically for medical treatment. Historical and autopsy studies arguably give a better, albeit different, insight into the natural history of atrioventricular septal defect. Berger pointed out that, without currently accepted treatment, 46% of the patients with an atrioventricular septal defect died in the first 6 months of life[17]. This group of patients could well have been missed in the study of Bull and her colleagues[5]. A second point is that the authors refer to the quality of life of these patients as "generally good" while it is known that most develop pulmonary vascular disease. Many will subsequently die in a cyanotic, polycythemic state with good chances for the known resulting disabling complications. The chances for a "generally good" quality of life, therefore,

seem to be rather slim for these patients. Thirdly, the operative mortality figures that are used by Bull and her colleagues[5] are outdated in the light of the current paper[3]. Finally, it is known that "patients with Down's syndrome have no predisposition to develop severe or precocious pulmonary vascular disease"[18]. In my opinion, therefore, patients with Down's syndrome and an atrioventricular septal defect should benefit from the same surgical care as all other patients.

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OVERALL CONCLUSIONS

The fact that the left atrioventricular valve in atrioventricular septal defect is trifoliate was observed by Peacock in 1846 and rediscovered by Carpentier in 1977. In the interval, the fashionable anatomical theory has been that the anterior leaflet of the otherwise normal mitral valve was "cleft" into two halves. The significance of this anatomical observation is that the "cleft" supposedly had to be closed at operation, but the result was not always favorable. Embryologically based nomenclature has, therefore, proven to be less than optimal. Additionally, this nomenclature is prone to instability, as names have to change with changing embryological concepts.

The results of operative treatment for patients with atrioventricular septal defect at our department are in keeping with the results that are reported worldwide. These results have improved over time and are now primarily dependant on the management of the left atrioventricular valve. The understanding of the function of this trifoliate valve is of paramount importance to the well being of the patient.

The precise configuration of the valve can be determined echocardiographically, and has a demonstrable impact upon its function. Surgical techniques should be guided by the morphometry determined by echocardiographic and direct observation. Furthermore, there is a morphometric relation between the configuration of the valve and that of the ventricular septum. This morphometric relation could very well be a reflection of a related embryological derivation.

The length of the narrow left ventricular outflow tract is also related to the atrioventricular valvar morphology. The usual systolic murmur in these patients often originates from the outflow tract, rather than being caused by left atrioventricular valvar regurgitation. The outflow tract constricts in all patients, and can be iatrogenically narrowed even more at primary repair or at replacement of the left atrioventricular valve. The surgical solution for this problem that has been developed in the course of this study is resection of the posterior wall of the outflow tract, thus suspending the valve prosthesis at the level of the aortic valve, rather than at the entrance to the outflow tract.

Down's syndrome should not be a contraindication for operation, because the quality of life without operation is undoubtedly worse if not unacceptable. But the quality of death may be an even more convincing argument in favour of operation, as unoperated children with Down syndrome have a high probability of dying of the extremely disagreeable sequels of obstructive pulmonary vascular disease.

SAMENVATTING

Een atrioventriculair septum defect met gescheiden kleporificia werd van oudsher een atrium septum defect van het "ostium primum" type genoemd. Deze conventie brengt ons meteen tot het probleem van de anatomische interpretatie, want relatief recent onderzoek heeft aangetoond dat het septum primum per se intact is. De reden voor de verwarring is begrijpelijk, omdat de atrioventriculaire kleppen bij deze afwijking naar apicaal zijn verplaatst en vastzitten aan de bovenrand van het ventriculaire septum. Dit inlet ventrikel septum defect is daarom effectief gelocaliseerd in het atrium. En omdat het defect "laag" in het atrium septum is gelocaliseerd schijnt het een defect te zijn van het embryologische primum septum dat deficient is. Anatomische nomenclatuur die gebaseerd is op embryologische percepties brengt daarom het risico met zich mee dat met veranderende embryologische inzichten, de nomenclatuur achterhaald raakt. De evolutie van de anatomische perceptie van 1846 tot heden is het onderwerp van hoofdstuk II.

Chirurgie voor atrioventriculair septum defect begon met "gesloten" (d.w.z. zonder hart-long machine) operaties, waarbij werd getracht het atrium septum defect te sluiten. De resultaten waren meestal onbevredigend, omdat deze gesloten techniek de chirurg niet in staat stelden het geleidingsweefsel te vermijden, dat vlakbij de rand van het defect loopt; terwijl pacemakers in die tijd nog science fiction waren, met name voor kinderen. Maar net zo belangrijk was het feit dat de gesloten techniek de chirurg niet in staat stelden de linker atrioventriculaire klep te behandelen, die vaak regurgiteert, wat vaak zelfs toeneemt na minder gelukkige chirurgie. De komst van open hart operaties gaf de chirurg de tijd die nodig is om deze afwijking zorgvuldig te opereren. De geschiedenis van de chirurgie van deze afwijking is het onderwerp van hoofdstuk III.

Het resultaat en de follow-up van de 133 operaties die in 20 jaar werden uitgevoerd voor atrioventriculair septum defect is onderwerp van hoofdstuk IV. Voor de lange termijn blijkt het functionele resultaat van de linker atrioventriculaire klep het belangrijkste te zijn. Bij het onderzoek van deze patienten viel ons op dat vele patienten een systolisch geruis vertoonden, dat we niet konden verklaren met regurgitatie van de linker atrioventriculaire klep. Deze bevinding was aanleiding een onderzoek te starten naar de vraag of de nauwe linker ventrikel uitstroombaan het geruis zou kunnen veroorzaken. We hebben dat echter niet kunnen aantonen. Wel bleek de uitstroombaan veelal nauwer te zijn dan de aorta. Bovendien contraheerde de uitstroombaan in alle gevallen. Deze bevindingen zijn onderwerp van hoofdstuk V.

Naar aanleiding van de echocardiografische studie werden vragen opgeworpen naar de preciese anatomie van de uitstroombaan. Daartoe werd een anatomisch

studie uitgevoerd, waarbij bleek dat de uitstroombaan het nauwst is in de atrioventriculaire septum defecten met gescheiden orificia. Tijdens deze studie werd een methode ontwikkeld om de achterzijde van de uitstroombaan te receseren bij vervanging van de atrioventriculaire klep door een prothese, om zo obstructie van de uitstroombaan door de prothese te voorkomen. Deze studie is het onderwerp van hoofdstuk VI.
Tijdens de echocardiografische studie van hoofdstuk V werd ook de linker atrioventriculaire klep gemeten en op functie beoordeeld. Het bleek dat de kleppen die regurgiteerden een andere configuratie hebben dan de kleppen die sufficient zijn. Zo zou het mogelijk moeten zijn om de groep die risico loopt op klepregurgitatie preoperatief al te identificeren. Hoofdstuk VII bevat de resultaten van dit deel van het onderzoek.
Hoofdstuk VIII betreft een studie die werd uitgevoerd in het Heart Museum van het Children's Hospital te Pittsburgh, U.S.A., naar de verhoudingen van het ventrikel septum en de relatie daarvan met de morfometrie van de linker atrioventriculaire klep. Resultaat van deze studie is een ontwikkelings sequentie van de linker atrioventriculaire klep. Bovendien blijken er zekere relaties tussen klep en septum te bestaan die wijzen in een gemeenschappelijke embryologisch deficiente derivatie.
Hoofdstuk IX tenslotte bevat enige visies op verschillende aspecten van de behandeling van kinderen met een atrioventriculair septum defect. Echocardiografie is tegenwoordig superieur in het afbeelden van de anatomische verhoudingen, zodat hartcatheterisatie veelal overbodig is. Een heel andere kwestie is de behandeling van kinderen met een Down syndroom, die frequent een atrioventriculair septum defect hebben. Met name in het Verenigd Koninkrijk gaan stemmen op om deze kinderen niet te opereren, omdat de levensverwachting niet verlengd zou worden. Tegen deze opvatting neem ik stelling, mede omdat de kwaliteit van leven voor deze kinderen belangrijk verbetert door een operatie. Zonder operatie is de kans op uiterst onaangename complicaties groot. Daarom wil ik er voor pleiten alle kinderen met deze afwijking te opereren.