THE RELATIONSHIP OF THE OUTLET SEPTUM TO THE AORTIC OUTFLOW TRACT IN HEARTS WITH INTERRUPTION OF THE AORTIC ARCH

We examined 13 hearts with concordant atrioventricular and ventriculoarterial connections and interruption of the aortic arch to establish and describe the morphologic features of the outflow tracts in relation to axial deviation and malalignment of the outlet septum as opposed to overriding of the arterial valvular orifices. Interruption in all cases but one was between the left common carotid and left subclavian arteries; the other arch was interrupted at the isthmus. A patent arterial duct and ventricular septal defect were universally present. When its borders were viewed from the right ventricle, the ventricular septal defect was perimembranous in seven hearts, had exclusively muscular borders in four hearts, and was doubly committed and juxta-arterial in the remaining two hearts. Malalignment between the muscular ventricular septum and outlet septum, or a fibrous raphe, as judged when the heart was viewed in its short axis, was found in 12 of the hearts. Posterior and leftward axial deviation of the outlet septum in its long axis was found in 4 of the 12 hearts and also in one heart that did not have short-axis malalignment. Attachments of the leaflets of the pulmonary valve in both right and left ventricles, however, were present in only one of the specimens, this being a case with a doubly committed and juxta-arterial defect. These separate features of the outflow tract in hearts with interruption of the aortic arch, therefore, require thorough assessment when surgical management is planned. All these variable features can be assessed preoperatively by cross-sectional echocardiography, which should be directed toward defining the degree of development and alignment of the outlet septum, as well as the length of the subpulmonary infundibulum. (*J THORAC CARDIOVASC SURG* 1995;109:1225-36)

Hala M. F. Al-Marsafawy, MD,* Siew Yen Ho, PhD, MRCPATH,
Andrew N. Redington, MD, FRCP, and Robert H. Anderson, MD, FRCPATH,
London, England

Interruption of the aortic arch is a rare congenital anomaly. It accounts for about 1% of all congenital heart diseases¹ and, if untreated, is usually rapidly fatal.² Although rare cases of isolated interruption have been reported, there is usually a per-

sistent arterial duct that supplies blood to the lower body and, most frequently, a ventricular septal defect (VSD) is also present. Interruption can also be associated with more complex lesions such as double-outlet right ventricle, common arterial trunk, aortopulmonary window, and discordant ventriculoarterial connections.^{3, 4}

The use of prostaglandin E₁ to maintain ductal patency has revolutionized the preoperative management of this condition, and most infants now survive to reach centers for tertiary care where surgical treatment can be undertaken.^{5, 6} There are increasing numbers of reports in the literature concerning the potential options for operation, and there is continuing debate as to the utility of single-versus two-stage repair. There can be little doubt that the single most important factor determining the outcome of operation is the morphology of the subaortic outflow tract, in particular the location and alignment of the outlet septum.

In this study, therefore, we examined the ana-

From the Department of Paediatrics, Royal Brompton Hospital and National Heart and Lung Institute and Hospital, London, England.

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Address for reprints: R. H. Anderson, MD, FRCPATH, Department of Paediatrics, National Heart and Lung Institute, Dovehouse St., London SW3 6LY, England.

*During this study, Dr. Al-Marsafawy was a Fellow in the Department of Paediatrics, Royal Brompton. Current address: Paediatric Cardiology Unit, Department of Paediatrics, Mansoura University Hospital, Mansoura, Egypt.

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Fig. 1. Diagram showing concept of septal malalignment and deviation as distinguished by viewing heart along its short axis (A) and long axis (B), respectively. Malalignment is present when outlet septum makes an angle (θ°) with muscular ventricular septum. Deviation is described when outlet septum tilts along its axis and protrudes into left or right ventricular outflow tract.

tomic variables that influence this morphology in a series of hearts with concordant segmental connections, concentrating on the anatomy of the VSD, the disposition of the outlet (infundibular or conal) septum, and the degree of overriding, if any, of the muscular ventricular septum by the arterial trunks.

Material and methods

We examined 13 hearts with interruption of the aortic arch taken from the cardiopathologic collection of the Department of Paediatrics, Royal Brompton National Heart and Lung Hospital. The arch was considered to be interrupted when there was neither actual nor potential continuity of flow from the ascending to the descending aorta (in other words, cases of acquired atresia with fibrous continuity between the arch and the descending aorta were excluded). The aortic arch of each heart was examined carefully and any variation in arterial pattern noted. The intracardiac anatomy of each heart was studied, with particular attention paid to the outlet (infundibular) septum, the subpulmonary infundibulum, the VSD, and any factor causing obstruction to flow of blood to the aorta. To describe fully the morphologic features of the

outflow tract, we must be precise with our terminology. We used the following definitions:

1. *Outlet (infundibular) septum* indicated the muscular structure separating the subaortic and subpulmonary outflow tracts.
2. *Subpulmonary infundibulum* indicated the free-standing sleeve of musculature supporting the semilunar attachments of the valvular leaflets and separating the cavity of the pulmonary outflow tract from the extra-cardiac space surrounding the aortic sinuses.
3. *Malalignment of the outlet septum* was deemed to exist when the plane of the outlet septum, as judged in the cardiac short axis, was not in the same line as the remainder of the muscular ventricular septum, also seen in the short axis (Fig. 1, A). Such septal malalignment is independent of the anatomic nature of the borders of the essential coexisting VSD. Thus malalignment of the outlet septum, or malalignment of a fibrous raphe between the arterial valves, can be found when the defect (viewed from the right ventricle) has a fibrous posteroinferior rim (perimembranous), has exclusively muscular borders, or is doubly committed and juxta-arterial (roofed by the conjoined leaflets of the aortic and pulmonary valves).

4. *Deviation of the outlet septum* was diagnosed when the long axis of the outlet septum was deviated either anteriorly (into the right ventricle) or posteriorly (into the left ventricle) relative to the long axis plane of the remainder of the muscular ventricular septum (Fig. 1, *B*).
5. *Overriding of an arterial valve* was diagnosed in the presence of biventricular attachments of the leaflets of one or the other, or both, of the arterial valves. The degree of overriding was quantified according to the ratio of the valvular circumference supported within each ventricle.

So as to establish any relation between the length of the subpulmonary infundibulum and the severity of obstruction to the aortic outflow tract, we measured the length of the subpulmonary infundibulum from the attachments of the bases of the pulmonary valvular leaflets to the lower end of the outlet septum above the VSD. Hegar dilators were used to measure the diameter of the aortic outflow tract, which was considered to be a circular structure. The length of the outlet septum on the left ventricular aspect was assessed in a similar manner to that used to quantify the subpulmonary infundibulum. Thus the shortest vertical distance was measured between the free edge of the outlet septum and the basal attachment of the leaflets of the aortic valve. The angle of malalignment between outlet and muscular ventricular septum, as seen in the short axis plane of the heart, was measured by viewing the heart from the left ventricular apex. Two cases were excluded from these measurements because the outlet septum was lacking.

Results

Of 13 hearts examined, 5 were from male and 7 from female infants. The sex was unknown in the other patient (the clinical data were taken from the medical records of those patients who were admitted to Royal Brompton Hospital from 1966 to 1993). The median age at death was 7 days.

Anatomy of the aortic arch. All arches were left-sided. The interruption was between the left subclavian and left common carotid arteries (so-called type B) in all cases but one, in which the interruption was distal to the left subclavian artery (so-called type A). An anomalous retroesophageal origin of the right subclavian artery was found in 4 of the 12 specimens with interruption between the left common carotid and the left subclavian arteries. The distal portion of each interrupted arch was connected to the pulmonary trunk via a patent arterial duct, which was the only source of blood supply to the lower body. There was a very low origin of the right pulmonary artery from the pulmonary trunk in one heart. The great arteries were normally related in all.

VSD. A VSD was present in all the hearts. The central fibrous body, incorporating fibrous continu-

ity between the leaflets of the aortic and tricuspid valves, formed the posterior border of the defect in eight hearts (perimembranous defects) (Fig. 2, *a*). In three, in contrast, the borders of the defect as seen from the right ventricle were entirely muscular (Fig. 2, *b*). The other two hearts had doubly committed and juxta-arterial defects, with fibrous continuity between the leaflets of the aortic and pulmonary valves forming the roof of the defect. When viewed from the right ventricular aspect, two perimembranous defects were seen to open to the inlet and another to the trabecular portion, whereas the other defects all opened between the ventricular outlets. One doubly committed and juxta-arterial defect was additionally perimembranous, whereas the other possessed a muscular posteroinferior border.

Anatomic substrates of obstruction to aortic flow.

Obstruction to the aortic outflow tract was evident in all cases. In the majority of cases, the anatomic substrates were related to an abnormal arrangement of the outlet septum. Malalignment of the outlet septum, or of the fibrous raphe between the arterial valves, as defined along the cardiac short axis, was present in 12 of 13 cases. These included the two hearts with doubly committed and juxta-arterial defects. When present, the outlet septum was hinged posteriorly to the area of tricuspid-aortic-mitral valvular fibrous continuity or to the posterior limb of the septomarginal trabeculation in 7 of 10 cases, all with outlet VSDs. The anterior component of the outlet septum then moved out of alignment with the muscular septum, to an extent between 15 and 35 degrees, and attached either to the anterolateral wall of the left ventricle or to an anomalous muscle bundle adherent to it (Fig. 3, *a* and *b*). This septal malalignment within the left ventricular outflow tract was not always obvious when the specimen was examined from the right ventricle (Fig. 4).

In two of the remaining cases with septal malalignment (not counting the two with absence of the outlet septum), the outlet septum was arranged in a more coronal plane, and was positioned at 40 and 80 degrees relative to the muscular ventricular septum. These hearts were associated with perimembranous defects that excavated to the trabecular and inlet components, respectively, when viewed from the right ventricle. In both hearts, the outlet septum was hinged to the back of the anterior limb of the septomarginal trabeculation and inserted to the anterolateral wall in the left ventricle (Fig. 5). Despite the septal malalignment, none of the nine cases with

Fig. 2. **a**, Right ventricular view of this heart shows VSD situated between anterior (*A*) and posterior (*P*) limbs of septomarginal trabeculation (*SMT*). This defect is described as perimembranous owing to fibrous continuity between tricuspid and mitral valves at its posteroinferior margin (*star*). Pulmonary valve arises exclusively from right ventricle. Outlet septum roofs VSD and passes to left ventricle. *Pulm.*, Pulmonary. **b**, This heart has interruption of aortic arch (*double-ended arrow*) between left common carotid and left subclavian arteries. VSD, viewed from right ventricle, has muscular posteroinferior (*post. inf.*) border. *Asc.*, Ascending; *Desc.*, descending. **c**, This heart, sectioned through arterial outflow tracts, shows doubly committed and juxta-arterial VSD with biventricular attachments of aortic valve. Outlet septum is absent. *Arrow* points to fibrous raphe between arterial valves. *Pulm.*, Pulmonary.

malaligned outlet septums had associated biventricular attachments (overriding) of the leaflets of either the pulmonary or aortic valve. The length of the subpulmonary infundibulum ranged from 3 to 6 mm in these hearts (Fig. 6, *a* and *c*). In contrast, the outlet septum when viewed from the left ventricle measured between 0.5 and 3 mm in length, and there was a gross disparity in the levels of attachment of the leaflets of the arterial valves (Fig. 6, *b*, *c*, and *d*). In one case, the outlet septum was a fibrous ridge.

Malalignment of the raphe between the arterial valves produced overriding of the aortic valve in one heart and overriding of the pulmonary valve in the other, in the two cases with doubly committed and juxta-arterial VSDs (Fig. 2, *c*, and Fig. 7).

Deviation of the outlet septum in its own long axis, additionally present in four of the nine hearts with septal malalignment, further narrowed the aortic outflow tract. The outlet septum was also deviated posteriorly in the one heart in which it was aligned with the muscular ventricular septal structures when viewed in

the short axis. Although the length of the outlet septum, as measured vertically from its free edge to the aortic valve, was less than 1 mm in this heart, it had an extensive oblique protrusion into the subvalvular area within the left ventricle (Fig. 8).

One heart had minimal malalignment (less than 5 degrees) without deviation of the outlet septum in its long axis. In this heart, obstruction to aortic outflow was the result of a prominent anterolateral muscle bundle together with anomalous attachments to the ventricular septum of an isolated cleft in the aortic leaflet of the mitral valve (Fig. 9).

An anomalous muscle bundle in the anterolateral wall of the left ventricle further aggravated obstruction to the subaortic outflow in another four hearts. The left ventricular free wall forming the anterior border of the outflow tract was hypertrophied in another six cases as a part of generalized left ventricular hypertrophy.

Associated abnormalities. The aortic valve had two leaflets in nine cases (69%), and the mean

Fig. 3. Two hearts showing septal malalignment. **a,** This left ventricular view shows anterior component of outlet septum malaligned and attached to anterolateral wall of left ventricle (*star*). **b,** Outlet septum in this heart is short and hinged to posterior margin (*star*) of VSD. Its anterior component, out of alignment with muscular ventricular septum, is attached to anomalous muscle bundle. *Ant. lat.,* Anterolateral.

Fig. 4. a, Right ventricular view of this heart shows VSD between limbs (anterior, *A*; posterior, *P*) of septomarginal trabeculation (*SMT*). Septal malalignment is not apparent in this view. *Double-ended arrow* indicates interruption of aortic arch between left common carotid and subclavian arteries. *Pulm.,* Pulmonary. **b,** This dissection of left ventricle of same heart shows outlet septum veering away from ventricular septum to be attached to anterolateral muscle bundle.

Fig. 5. **a**, Right ventricular view of this heart shows perimembranous VSD extending apically toward trabecular portion. Diminutive outlet septum is hinged to anterior limb (*A*) of septomarginal trabeculation (*SMT*). *P*, posterior; *Pulm.*, pulmonary. **b**, Outlet septum (arrows) inserts into anterolateral wall (star) in left ventricle. Posteroinferior margin (triangle) of VSD is area of fibrous continuity between tricuspid, aortic, and mitral valves. Aortic valve has two leaflets.

diameter of the valve was 4.6 mm. The left ventricular outflow tract narrowed to 2 mm in diameter in three of those cases, and poststenotic dilation of the aorta above a bicuspid valve was seen in another. Other associated malformations are listed in Table I.

Measurements of subpulmonary infundibulum and aortic outflow tract. Measurements were taken in 11 hearts (the two hearts with doubly committed and juxta-arterial VSD were excluded). The mean length of the subpulmonary infundibulum was 4.95 mm (standard deviation ± 1.9), and the mean diameter of the aortic outflow tract was 2.8 mm (standard deviation ± 0.75). A coefficient of 0.25 was found for the correlation of these values ($p = 0.46$, not significant).

Discussion

Interruption of the aortic arch is a rare congenital anomaly. Nearly all patients with this lesion are seen as neonates with severe intractable heart failure and profound metabolic acidosis.⁷ Timely infusion of prostaglandin E₁ now allows maintenance of patency of the arterial duct and improves perfusion to the lower body, thereby improving renal function

and diminishing acidosis.^{5,6} Without surgical intervention, nonetheless, nearly all patients die in the first year of life.⁸

In our autopsy series, interruption in the aortic arch was most commonly found between the left common carotid and left subclavian arteries, with only one heart having interruption at the isthmus. The arterial duct was patent in all our cases and supplied blood to the descending aorta and the lower body. This pattern approximates that seen in life.⁹ A typical spectrum of associated cardiovascular lesions similar to those found in life⁹ was also present in our series.

In normal hearts, the outlet septum, albeit of insignificant size, inserts between the anterior and posterior limbs of the septomarginal trabeculation. In the majority of our cases with interruption of the aortic arch, the outlet septum was a seemingly prominent structure that was malaligned when viewed in the cardiac short axis relative to the remainder of the ventricular septum. In this study septal malalignment in the short axis is distinguished from deviation of the outlet septum in its long axis to define more clearly the anatomic substrate to

Fig. 6. Two hearts showing differing degrees of development of outlet septum. **a**, This right ventricular view of first heart shows diminished subpulmonary (*Subpulm.*) infundibulum at roof of VSD. *Broken line* marks attachments of leaflet of pulmonary (*Pulm.*) valve. **b**, Same heart, viewed from left, shows how small outlet septum is. In this display, outflow tracts are distorted such that pulmonary (*Pulm.*) valve is visible through VSD. However, view in **a** clearly shows no override of pulmonary valve across septum. **c**, Second heart shows extensive subpulmonary (*Subpulm.*) infundibulum with attachments of pulmonary valve solely to right ventricle. Outlet septum (*star*) also appears extensive. **d**, When sectioned through outflow tracts, much of "outlet septum" is in reality musculature of subpulmonary infundibulum (*double-ended arrow*). Outlet septum (*brace*) when measured from its free edge to attachment of leaflets of aortic valve (*broken line*) is relatively short. *Pulm.*, Pulmonary.

Fig. 7. This section through heart with doubly committed and juxta-arterial VSD shows override of pulmonary valve across crest (*triangle*) of defect. Posteroinferior border of defect is muscular (*star*). In absence of outlet septum, raphe (*arrow*) between aortic and pulmonary (*Pulm.*) valves is malaligned into left ventricle.

obstruction. Each feature is an independent variable, and each can be recognized and distinguished on orthogonal views of cross-sectional echocardiograms.

Malalignment of the septal structures resulted in a large VSD, a universal feature of our cases, but a defect that was observed to show variability in its anatomic borders. In previous anatomic studies of interruption of the aortic arch,^{1, 4, 10-12} such VSDs have usually been described simply as representing the "malalignment type." Because the outlet septum occupies a different plane from the remaining muscular components of the ventricular septum, this description is undoubtedly accurate. However, it should not then be presumed that "malalignment type" gives a full description of the salient anatomy of the defect. As our present study shows, this is far from the case. Thus one referee of the initial draft of this paper commented as follows: "that most of the ventricular septal defects are described as being both of the perimembranous type and of the malalignment type may seem inconsistent to adherents of other schools of nomenclature." Indeed! It re-

mains a fact that it is necessary to describe the anatomic nature of the borders of the defect (perimembranous, muscular, or doubly committed and juxta-arterial) in addition to the associated feature of septal malalignment.

This is more so the case because there are several types of VSD associated with septal malalignment. For example, hearts with straddling and overriding of the tricuspid valve are characterized by malalignment between the atrial and ventricular septal structures, whereas hearts with discordant ventriculoarterial connections can have the deviated outlet septum located within the right ventricle. The particular feature of the malalignment seen in our series was such that the subsequent location of the malpositioned outlet septum compromised the flow into the aortic outflow tract. This arrangement, of course, is well known. It is usually stated, however, that, in addition, the pulmonary valvular orifice overrides the ventricular septum in association with biventricular origin of the leaflets of the pulmonary valve.^{13, 14}

On close examination, none of our cases, having excluded the two in which the outlet septum was absent, had overriding of the pulmonary valve despite the unequivocal short axis malalignment of the outlet septum. We suggest that this is the consequence of the length of the free-standing subpulmonary infundibulum. When there is a relatively long subpulmonary infundibulum, the pulmonary valvular orifice is carried away from the crest of the ventricular septum. Hence the lower margin of the infundibulum is in line with the crest of the muscular septum, and there is no perceived overriding. In contrast, when the subpulmonary infundibulum is lacking, as in the cases with doubly committed and juxta-arterial defects, the pulmonary valvular orifice is much closer to the crest of the septum, and its leaflets are then attached in both ventricles.

It has previously been accepted that overriding of the pulmonary trunk favors flow of blood into the pulmonary trunk at the expense of the preductal aorta, and this is manifest by the small aortic root and hypoplasia or interruption, or both, of the aortic arch.^{15, 16} Our data failed to demonstrate, however, any linear correlation between either the length of the subpulmonary infundibulum or the degree of overriding of the pulmonary valvular orifice and the diameter of the aortic outflow tract. It is perhaps worthy of comment in this context that in the normal heart the orifice of an arterial valve cannot override the ventricular septum, inasmuch as when the ven-

Fig. 8. Outlet septum in this heart is in alignment with rest of ventricular septum but its inferior part (*arrowheads*) protrudes into aortic outflow tract. Pulmonary valve (*dotted line*) is attached at considerably higher level than aortic valve (*broken line*). *Double-ended arrow* indicates long subpulmonary infundibulum and *brace* marks true length of outlet septum. *Pulm.*, Pulmonary.

tricular septum is intact it is impossible to have biventricular origin of the arterial valvular leaflets. Although this feature has been said by Kleinert and Geva¹⁷ to represent valvular overriding, it is more properly designated as overriding of the sinuses of the arterial trunk. Additional factors that may aggravate the obstruction to the systemic blood flow were also encountered in our hearts. A bicuspid aortic valve was present in nine hearts, but this feature in itself may not cause stenosis. An anterolateral muscle bundle¹⁸ of the left ventricle was present in five cases and hypertrophy of the anterolateral wall of the left ventricle was prominent in six cases.

With the increasing recognition that operation can be successful in infants with interrupted aortic arch, it becomes necessary to determine the optimal surgical approach.¹⁹⁻²² Crucial among the decisions to be made is whether to attempt excision or resection of the muscular outlet septum. Decisions of this nature cannot be made exclusively on the basis of examination of an autopsied series of hearts, which of necessity views the worst end of the anatomic spectrum. It is also highly pertinent that in the recent multiinstitutional study reported by Jonas and colleagues²³ fewer than 20% of patients were identified clinically to have important subaortic obstruction. Furthermore, optimal outcome was more often the result of appropriate relief of obstruction

Fig. 9. There is near alignment of septal structures but anomalous attachments of cleft of mitral valve together with anterolateral muscle bundle obstruction of aortic outflow. Tear in area of anomalous attachment (*small arrows*) is artifactual.

Table I. Morphologic features in 13 hearts with interruption of aortic arch

No.	Age in days	Sex	VSD type	VSD size (mm)	Ao override	Ao valve (diameter in mm)	Pulm. override	Pulm. valve	Malalignment of outlet septum	Deviation of outlet septum	Other anatomic substrates of obstruction to aortic blood flow
Interruption at isthmus											
1	464	F	Perim. inlet	5 × 7	—	2 leaflets raphe (6)	—	3 leaflets	+ <5°	—	Antlat. muscle bundle, acces. cords of cleft MV attached to septum
Interruption between left common carotid and left subclavian arteries											
2	4	F	Perim. outlet	6 × 8	—	2 leaflets (4)	—	3 leaflets	+ 20°	+	Hyper. antlat. wall
3	3	F	Perim. outlet	5 × 6	—	3 leaflets (4.5)	—	3 leaflets	+ 25°	—	Antlat. muscle bundle
4	6	M	Perim. outlet	5 × 5	—	2 leaflets (5)	—	3 leaflets	+ 15°	+	Hyper. antlat. wall
5	3	F	Perim. outlet	5 × 6	—	2 leaflets (3.5)	—	3 leaflets	+ 20°	—	Hyper. antlat. wall
6	3	M	Perim. outlet	3 × 5	—	2 leaflets (4)	—	3 leaflets	+ 35°	—	Hyper. antlat. wall
7	8	M	Musc. outlet	6 × 7	—	2 leaflets (6)	—	3 leaflets	+ 10°	+	Hyper. antlat. wall
8	27	M	Musc. outlet	5 × 7	—	2 leaflets (5)	—	3 leaflets	+ 15°	+	Antlat. muscle bundle
9	6	F	Perim. trab.	5 × 5	—	2 leaflets (4)	—	3 leaflets	+ 40°	—	Antlat. muscle bundle
10	14	F	Perim. inlet	7 × 8	—	2 leaflets (4)	—	3 leaflets	+ 80°	—	Hyper. antlat. wall
11	48	F	Musc. outlet	5 × 6	—	3 leaflets (4)	—	3 leaflets	—	+	—
12	—	—	Musc. DCJA	4 × 5	—	3 leaflets (5)	+	3 leaflets	+ 20°	—	—
13	13	M	Perim. DCJA	4 × 6	+	3 leaflets (4.5)	—	3 leaflets	+ 30°	—	Antlat. muscle bundle

Ao, Aorta; *pulm.*, pulmonary; *LV*, left ventricle; *subpulm.*, subpulmonary; *F*, female; *perim.*, perimembranous; *antlat.*, anterolateral; *acces.*, accessory; *MV*, mitral valve; *PAD*, patent arterial duct; *POF*, probe patent oval fossa; *LSCV*, left superior caval vein; *CS*, coronary sinus; *hyper.*, hypertrophied; *retroesoph.*, retroesophageal; *RSCA*, retroesophageal right subclavian artery; *R*, right; *M*, male; *trab.*, trabeculation; *musc.*, muscular; *OFD*, oval fossa defect; *DCJA*, doubly committed and juxta-arterial.

at the level of the aortic arch, and surgical attempts to relieve subaortic muscular obstruction carried a high mortality. It remains an anatomic fact, nonetheless, that resection of the outlet septum is a feasible option, because this part of the septum never contains any conduction tissues. The problem is how, and when, to approach the septum. The frequent small size of both the ascending aorta and the aortic valvular orifice makes difficult transaortic resection of the obstructing muscle.¹⁹ Freedom and colleagues¹¹ considered the possibility of approaching the obstruction through the VSD, and this was successfully achieved by those who used a transatrial approach to the VSD and the subaortic area.^{9,24} More recently, Watanabe and associates²⁵ have avoided the need for resection by placing traction sutures on the outlet septum during closure of the VSD and pulling it toward the right ventricular side.

Irrespective of whether the problem of the muscular subaortic obstruction should be dealt with at

the time of initial repair, it is advantageous to identify deviation of the outlet septum, because it is patients with this additional feature who are the most obvious candidates for the more aggressive option of relieving potential obstruction to the flow of blood into the aorta. In this regard, we caution that although the outlet septum may seem to be an extensive structure in the right ventricle, much of the perceived "septum" may be the free-standing subpulmonary infundibulum. From the left ventricular aspect, the outlet septal musculature, as in nine of our hearts, may extend no more than 2 mm from the lower attachments of the aortic valvular leaflets. In these cases, resection of the anterolateral musculature or bundle may be a safer option.

There is a need, therefore, to define preoperative criteria to predict accurately those cases likely to have significant early subaortic obstruction. Previously accepted angiographic criteria have suggested

Associated malformations	Length of outlet septum in LV (mm)	Length of subpulm. infundibulum (mm)	Diameter of LV outflow tract (mm)
PAD, POF, LSCV to CS, poststenotic dila- tion aorta	5	9	3
PAD, POF retro- esoph. RSCA	3	5	3
PAD, POF very low origin of R pulm. artery	2	4	3
PAD, POF			
PAD, POF	1	6	2
PAD, POF	1(fibrous ridge)	4	2
PAD, POF	1	3.5	2
PAD, POF	<1	4	4
PAD, POF retro- esoph. RSCA	2	4	4
PAD, POF retro- esoph. RSCA	0.5	3	3
PAD, OFD retro- esoph. RSCA	1	4	3
PAD, POF	<1	8	2
PAD, POF	—	—	—
PAD, OFD	—	—	—

a systolic ratio of the diameter of the left ventricular outflow tract and that of the descending aorta at the level of the diaphragm of 0.6 or less.²⁶ Bove and associates²⁷ suggested the use of a ratio of 0.7 between the diastolic diameter of the left ventricular outflow tract and that of the descending aorta at the level of the diaphragm. The cross-sectional area of the left ventricular outflow tract indexed to body surface area has also been proposed by other investigators as a more accurate means of identifying patients with significant subaortic obstruction.²⁸ All of these are indirect criteria. Our study shows that it is equally important carefully to differentiate echocardiographically between malalignment and axial deviation of the outlet septum, as well as to demonstrate the extent of the outlet septum below the basal attachment of the leaflets of the aortic valve. This can give clues as to whether transatrial (well-developed outlet septum) or transpulmonary (hypoplastic outlet septum) resection can potentially be undertaken, together with how much of the outlet septum can safely be resected without the risk of damaging the aortic valve. The clinical indications of our study, therefore, are clear. It should be possible,

with the use of cross-sectional echocardiography, to examine all the morphologic variables that we have described. The degree of development, malalignment, and deviation of the outlet septum; the length of the subpulmonary infundibulum; and the presence of overriding of one or the other of the arterial trunks can and should be carefully defined, inasmuch as they are critical to subsequent optimal surgical management.

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