

Double-Outlet Right Ventricle: Morphologic Demonstration Using Nuclear Magnetic Resonance Imaging

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Sixteen patients with double-outlet right ventricle, aged 1 week to 29 years (median 5 months), were studied with a 1.5 tesla nuclear magnetic resonance (NMR) imaging scanner. Two-dimensional echocardiography was performed in all patients. Thirteen patients underwent angiography, including nine who underwent subsequent surgical correction. Three patients underwent postmortem examination.

Small children and infants were scanned inside a 32 cm diameter proton head coil. Multiple 5 mm thick sections separated by 0.5 mm and gated to the patient's electrocardiogram were acquired with a spin-echo sequence and an echo time of 30 ms. A combination of standard and oblique imaging planes was used. Imaging times were <90 min. The NMR images were technically unsuitable in one patient because of excessive motion artifact.

In the remaining patients, the diagnosis of double outlet right ventricle was confirmed and correlated with surgical and postmor-

tem findings. The NMR images were particularly valuable in demonstrating the interrelations between the great arteries and the anatomy of the outlet septum and the spatial relations between the ventricular septal defect and the great arteries. Although the atrioventricular (AV) valves were not consistently demonstrated, NMR imaging in two patients identified abnormalities of the mitral valve that were not seen with two-dimensional echocardiography. In one patient who had a superoinferior arrangement of the ventricles, NMR imaging was the most useful imaging technique for demonstrating the anatomy.

In patients with double-outlet right ventricle, NMR imaging can provide clinically relevant and accurate morphologic information that may contribute to future improvement in patient management.

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Double-outlet right ventricle describes a specific type of ventriculoarterial (VA) connection in which both great arteries are connected to the morphologic right ventricle. In this context, a great artery is considered to be connected to a ventricle when >50% of its circumference (measured at the level of the semilunar valve) originates from that ventricle (1). Patients with this diagnosis are unlikely to form a single clinical entity. Although they share a specific type of connection, this may occur in conjunction with any atrial arrangement and with any type or mode of atrioventricular (AV) connection. In addition, the condition is usually associated with a range of cardiac malformations that almost always includes the presence of a ventricular septal defect (2,3).

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The treatment of patients with this type of connection is complex and requires an accurate understanding of the individual cardiac morphology (4-9). Precise presurgical evaluation and correct surgical selection are important determinants of prognosis. Although it is always preferable to consider the anatomy of each patient individually, several morphologic studies (3,7-10) have identified specific features common to all groups of patients with double-outlet right ventricle that can be used as a basis for making a surgical decision. Imaging methods currently available for providing this information include two-dimensional echocardiography and angiography, both of which are accurate but have important limitations (10-13). The purpose of our study, therefore, was to evaluate the clinical value of another imaging technique, nuclear magnetic resonance (NMR) imaging, in demonstrating cardiac anatomy in patients with double outlet right ventricle.

Methods

Study patients (Table 1). Between March 1987 and August 1989, all patients at Guy's Hospital with the diagnosis of double-outlet right ventricle and concordant AV connections

Table 1. Clinical and Imaging Data in 16 Patients

Pt No.	Age	Associated Anatomy	Angio	Corrective Surgery	PM	NMRI Findings
1	4 mo	Blalock-Taussig shunt	Yes	—	—	Agreed
2	11 wk	—	Yes	—	Yes	Agreed
3	5 yr	Banding of PT	Yes	Yes	—	Agreed
4	17 mo	Banding of PT	Yes	Yes	—	Agreed
5	29 mo	Banding of PT	Yes	Yes	Yes	Agreed
6	18 mo	Banding of PT	Yes	Yes	—	Fibromuscular subaortic stenosis seen with NMRI, confirmed at surgery
7	3 wk	Tetralogy of Fallot	Yes	Yes	—	Agreed
8	11 mo	Tetralogy of Fallot	Yes	Yes	Yes	Agreed
9	1 wk	—	—	—	—	Motion artifact
10	8 yr	Left atrial isomerism, aortic hypoplasia, bilateral PA bands	Yes	—	—	Agreed
11	5 wk	—	Yes	Yes	Yes	Agreed
12	6 wk	Superoinferior ventricular arrangement, straddling, MV	—	—	—	Ventricular arrangement and straddling MV seen with NMRI but not with echo
13	2 wk	AVSD, hypoplastic PA, Blalock-Taussig shunt	—	—	—	Agreed
14	5 mo	Restrictive VSD	Yes	Yes	—	MV involvement with VSD seen with NMRI but not with echo
15	29 yr	PVD	Yes	—	—	Agreed
16	9 yr	Banding of PT	Yes	—	—	Agreed

Angio = angiography; AVSD = atrioventricular septal defect; echo = two-dimensional echocardiography; NMRI = nuclear magnetic resonance imaging; MV = mitral valve; PA = pulmonary artery; PM = postmortem examination; Pt = patient; PT = pulmonary trunk; PVD = pulmonary vascular disease; VSD = ventricular septal defect.

confirmed by two-dimensional echocardiography were entered into the study. Sixteen patients were examined; their ages ranged from 1 week to 29 years (median 5 months). Associated malformations included aortic coarctation in two patients, AV septal defect associated with hypoplasia of the pulmonary arteries in one patient and isomerism of the left atrial appendages in one. At the time of the study, four patients had undergone palliative surgery, with banding of the pulmonary trunk in two and a modified Blalock-Taussig shunt in two. Angiography was performed in 13 patients and 9 of these subsequently underwent surgical correction. Four patients died; postmortem examination was performed in three.

Nuclear magnetic resonance (NMR) imaging. The Ethical Committee of Guy's Hospital approved the use of NMR imaging for examining children with congenital heart disease in 1987. Full informed consent was obtained from the parents of each child entered into the study.

Images were obtained with a 1.5 tesla whole body NMR imaging system (Philips Gyroscan). Infants and small children were positioned inside a standard 32 cm diameter proton head coil. Sedation with chloral hydrate (50 to 75 mg/kg body weight) was used in infants and small children. Simultaneous sections, 5 mm thick, separated by 0.5 mm and gated to the patient's electrocardiogram (ECG), were acquired with use of a T₁-weighted, spin-echo sequence with an echo time of 30 ms. Although the matrix size

was 256 × 256, only 180 signals were used to construct the final image. This meant that the time of scanning could be reduced without unduly affecting the resolution of the final image. The field of view was 200 to 250 mm. Two signal averages were performed for each examination and a typical scanning period for a series of seven sections was approximately 6 min (14). Sections were acquired in orthogonal and oblique imaging planes. Their selection was made by a cardiologist who was present during each examination and was familiar with the clinical and echocardiographic findings.

In all cases, an initial series of sections were taken in a transverse plane, encompassing structures between the base of the heart and the aortic arch. The connections and relative positions of the great arteries were then determined from these images.

The choice of subsequent imaging planes depended on the anatomy displayed from these initial images. Particularly useful planes included an oblique coronal plane rotated around a superoinferior axis and directed along the plane of the ventricular septum to demonstrate the ventricular septal defect and an imaging plane directed at right angles to the outlet septum, either oblique sagittal or oblique coronal, rotated around a superoinferior axis to demonstrate the anatomy of the infundibulum (Fig. 1). The total period of scanning for the complete examination varied with the number of imaging planes acquired but was always <90 min.

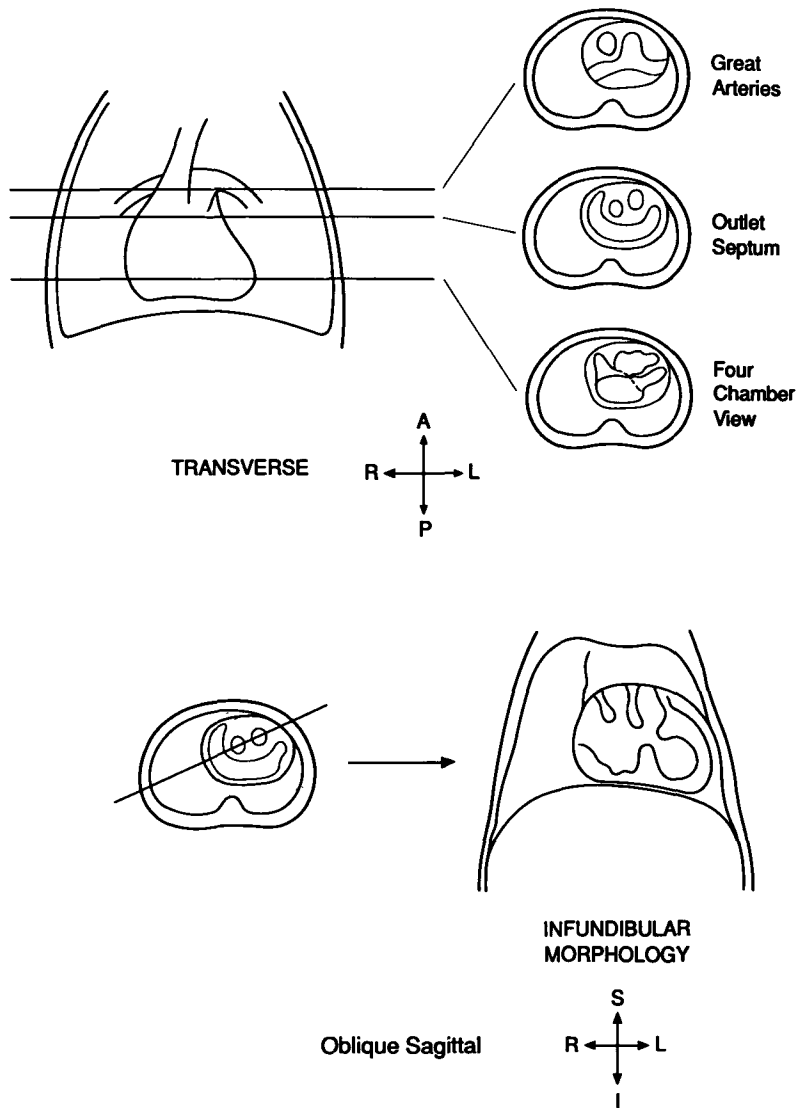


Figure 1. Some of the more frequently used imaging planes. Sections from transverse imaging planes (**top**) demonstrate the outlet septum and the interrelations of the great arteries. An oblique sagittal plane (**bottom**) directed through the outflow tracts demonstrates the morphology of the infundibulum. A = anterior; I = inferior; L = left; P = posterior; R = right; S = superior.

Anatomic criteria for determining the surgical technique. Previous studies (3,7-10) have shown that patients with double-outlet right ventricle may have specific anatomic features that can be used as criteria for predicting the optimal surgical technique. A retrospective analysis of the NMR images was performed to determine whether such criteria could be identified.

Anatomic specimens. Hearts from patients in our series who underwent postmortem examination were dissected to display the anatomy as it would be seen in cross section from imaging planes used during NMR imaging. The anatomic specimens were displayed for comparative purposes alongside corresponding NMR images. A heart from the Cardio-pathological Museum of the National Heart and Lung Institute has also been used for comparison in one case (Fig. 5B).

Results

Image quality. The NMR images were of diagnostic quality in 15 of the 16 patients. In one patient, motion artifact

occurring after inadequate sedation resulted in technically poor images that were of limited diagnostic value.

Accuracy of NMR images. Nine patients subsequently underwent corrective surgery and one patient who died soon after palliative surgery underwent postmortem examination. The accuracy of the NMR imaging findings was confirmed in these 10 patients. Three other patients had additional angiography and all 16 underwent two-dimensional echocardiography. There were no major discrepancies between the findings from these investigations and the NMR images. Confirmation of double-outlet right ventricle was made with NMR imaging in the 15 patients with technically satisfactory images from a series of sections taken in the transverse imaging plane. By projecting sections that demonstrated the great arteries onto more inferior sections that demonstrated the ventricles and ventricular septum, it was possible to assess accurately the proportional origin of a great artery from a particular ventricle, thereby determining its connection. In this way, double outlet right ventricle was differentiated from discordant and concordant VA connections (Fig. 2).

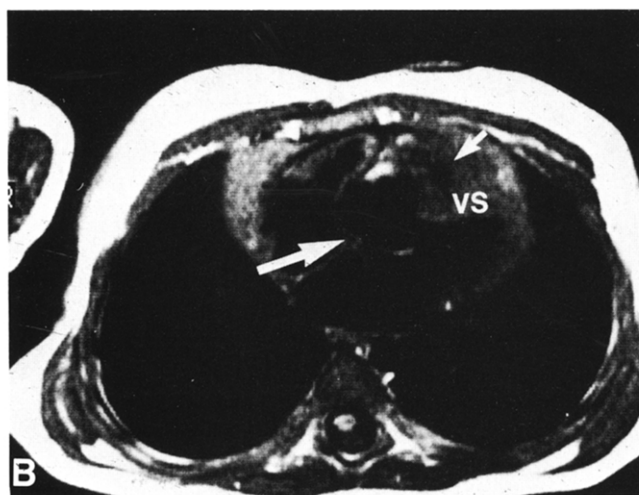
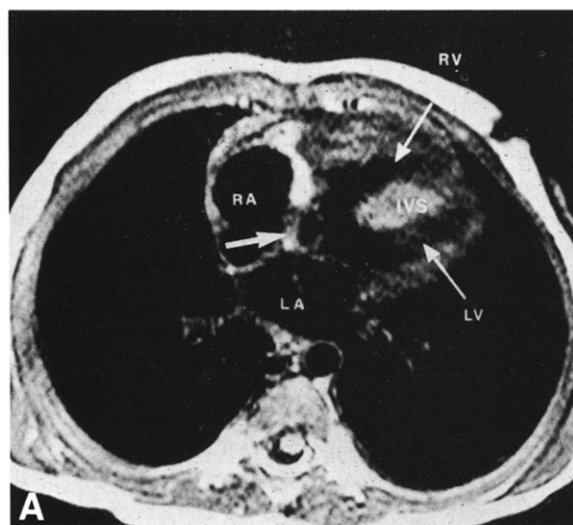


Figure 2. Sections from transverse planes demonstrating how the ventricular connection of a great artery (in both cases, the aorta identified at the level of the valvular leaflets) can be determined. **A**, The aorta (large arrow) has <50% of its circumference connected to the right ventricle (RV) in a patient (not in this series) with concordant ventriculoarterial connections. **B**, Patient 7. The aorta (large arrow) has >50% of its circumference connected to the right ventricle (small arrow) in a patient with double-outlet right ventricle. IVS = interventricular septum; LA = left atrium; LV = left ventricle; RA = right atrium; VS = ventricular septum.

Anatomic Features of Surgical Importance

Previous morphologic studies (3,7-10) identified several important anatomic features that are directly relevant to the surgical management.

Great arteries. In 15 of the 16 patients, the anatomic interrelation between the great arteries was clearly established (Fig. 3). In one patient, NMR images were nondiagnostic. The aorta in relation to the pulmonary trunk was posterior and to the right (Fig. 3A) in eight patients (similar to the arrangement in the normal heart), anterior and to the right in four (Fig. 3C) and anterior and to the left in three (Fig. 3E). The aortic arch was identified to the left of the

spine in 14 patients and was to the right in 1 patient. Hypoplasia of the pulmonary trunk and arteries was identified in one patient and hypoplasia of the thoracic aorta was seen in another. In three patients who underwent palliative surgical banding of the pulmonary trunk, excellent views of the pulmonary trunk and its branches were obtained; distortion of the pulmonary arteries was not seen.

In all cases, sections taken from transverse imaging planes were of greatest value. Sections from sagittal planes were also useful in determining the relative positions of the great arteries. These arteries were identified by following serial sections and observing the progression of the aortic arch from the ascending aorta and the branching into separate pulmonary arteries from the pulmonary trunk (Fig. 3A). In eight patients, the great arteries were seen to spiral around each other with interrelations similar to those seen in the normal heart (Fig. 3A). In the remaining patients, the great arterial trunks ascended parallel to each other.

Outlet septum. The presence and orientation of the outlet septum were readily assessed from images obtained in a transverse plane. Images from oblique coronal and oblique sagittal planes were also useful and were of particular value in assessing the presence of subvalvular stenosis. The NMR images were nondiagnostic in one patient. The orientation of the outlet septum varied according to the interrelations between the great arteries (Fig. 3). In eight patients whose great arteries were related in a fashion similar to that in the normal heart the outlet septum was at right angles to the ventricular septum and, except in the patient with a superoinferior arrangement of the ventricles, was seen to fuse with the anterior limb of the septomarginal trabeculation (Fig. 3B). Subvalvular aortic stenosis (which had developed after banding of the pulmonary trunk) was demonstrated clearly in one of these patients. It was excluded in the other seven patients. Subvalvular pulmonary stenosis was evident in one patient with tetralogy of Fallot, but with the aorta predominantly connected to the right ventricle (Fig. 4). Appropriate imaging sections were not acquired in one patient; stenosis was excluded in the other six patients.

In four patients whose aorta was anterior and to the right of the pulmonary trunk, the outlet septum was observed to be parallel to the plane of the ventricular septum, fusing with the ventriculoinfundibular fold (Fig. 3D). Subvalvular pulmonary stenosis was clearly demonstrated in two patients and excluded in two. In the remaining three patients, whose aorta was anterior and to the left of the pulmonary trunk, the outlet septum was seen to be hypoplastic in one patient and absent as a muscular structure in another (making the defect doubly committed in both). In the third patient, the outlet septum was observed to be at right angles to the ventricular septum, fusing with the septomarginal trabeculation (Fig. 3F). Subvalvular stenosis was not identified in any of these patients.

Ventricular septal defect. The NMR images were of poor quality in one patient and the ventricular septal defect was not visualized. One patient had an associated deficiency of

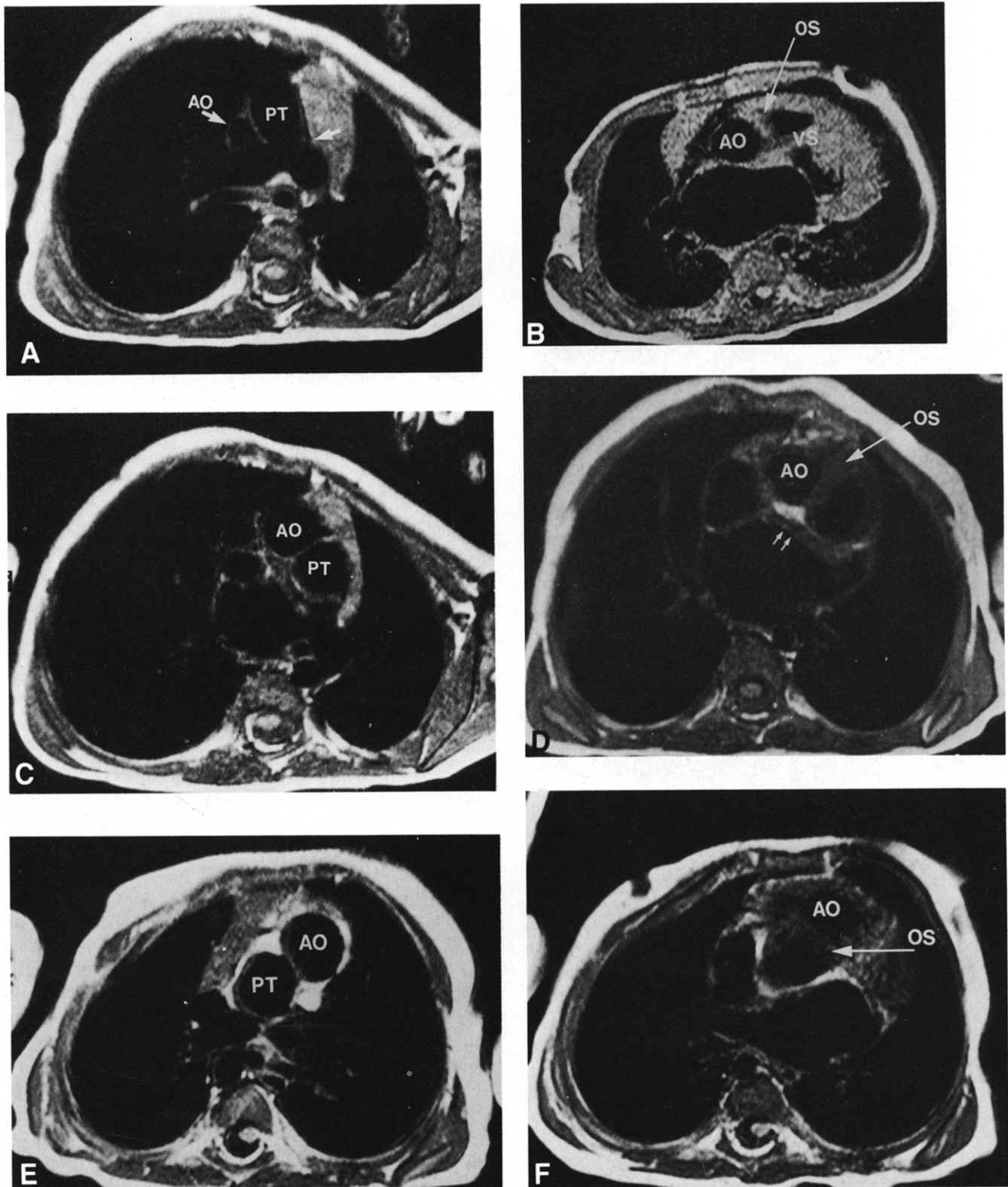


Figure 3. Sections from transverse planes taken through the great arteries together with corresponding sections from the same patients taken through the outflow tracts. **A**, Patient 14. This patient has a great arterial arrangement similar to the normal heart. The pulmonary trunk (PT) is identified with its bifurcation (arrow) into separate pulmonary arteries and is seen to spiral around the aorta (AO), which is posterior and to the right. **B**, In the same patient, the outlet septum (OS) is at right angles to the ventricular septum (VS) and

fuses with the anterior limb of the septomarginal trabeculation. **C**, Patient 4. In this patient, the aorta (AO) is anterior and to the right of the pulmonary trunk (PT) and the outlet septum (D) is now parallel to the plane of the infundibular fold (small arrows). **E**, Patient 5. In this patient, the aorta (AO) is now anterior and to the left of the pulmonary trunk (PT) and the outlet septum (OS) (F) is at right angles to the ventricular septum and fuses with septomarginal trabeculation as in **B**.

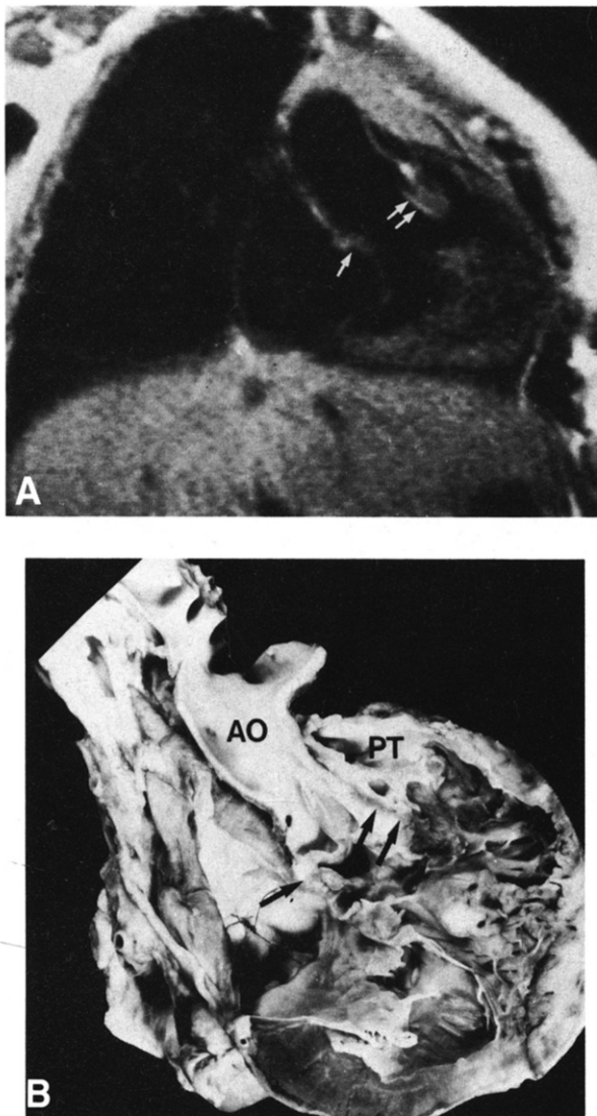


Figure 4. Patient 8. **A**, Section taken in an oblique sagittal plane from a patient with double-outlet right ventricle and tetralogy of Fallot. There is fibrous continuity between the valvular leaflets of the tricuspid and aortic valves (single arrow). A single subpulmonary infundibulum (double arrows) with subpulmonary stenosis is also demonstrated. **B**, Postmortem specimen from the same patient. Abbreviations as in Figure 3; arrows identified as in A.

AV septation. The ventricular septal defect was identified in the remaining 14 patients from a variety of views, including transverse, sagittal and oblique sagittal imaging planes. By using the criteria described by Baker et al. (15), it was subsequently possible in 12 patients to determine whether the posteroinferior rim of the ventricular septal defect was bordered by muscle (Fig. 5) or by the central fibrous body. In two patients, it was not possible to determine whether the margins were muscular or composed of fibrous continuity between the leaflets of the AV valves. In 14 patients, the ventricular septal defect was seen to open into the outlet components of the right ventricle. No defects were identified

within the apical trabecular component of the muscular septum.

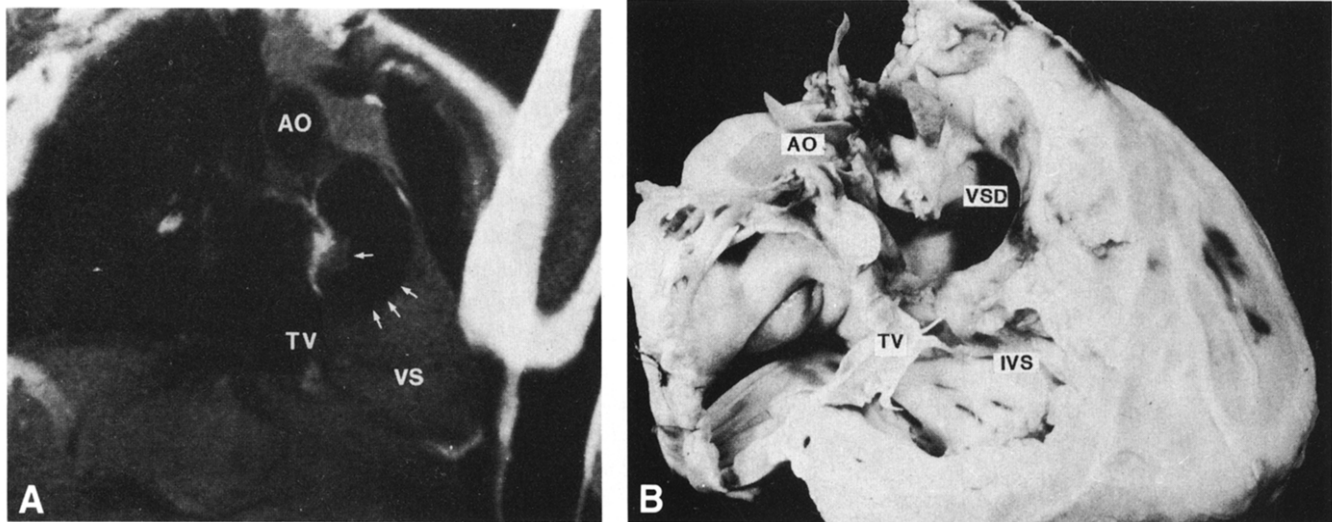
The roof of the ventricular septal defect was identified in 15 patients. This was formed by a leaflet of one arterial valve in 12 patients (aortic in 5, pulmonary in 7); in 1 patient, it was the ventriculoinfundibular fold, the defect then opening to the subaortic infundibulum. In the other two patients, it was roofed by fibrous continuity between the leaflets of the arterial valves in one and by a grossly hypoplastic muscular outlet septum in the other, both defects being doubly committed and immediately juxtaarterial. One patient had clinical features suggesting spontaneous closure of the ventricular septal defect. In this case, tension apparatus of the mitral valve was identified from a variety of views as being attached to borders of the ventricular septal defect (Fig. 6).

Infundibular morphology. The infundibular morphology was demonstrated in 13 patients from images taken in a variety of planes, including transverse, oblique coronal, through the plane of both great arteries (Fig. 1) and oblique sagittal, along the plane of the ventricular septum. No comment could be made in three patients whose NMR images were of poor quality or had not been acquired in the appropriate planes.

A single subpulmonary infundibulum was identified in five patients with fibrous continuity between the leaflets of the tricuspid and aortic valves (Fig. 4). A single subaortic infundibulum was demonstrated in three patients. In two of these patients, fibrous continuity was seen between the leaflets of the tricuspid and pulmonary valves. In the other patient, these valvular leaflets were separated by muscle, but the ventriculoinfundibular fold was poorly developed. Bilateral infundibulums were identified in three patients (Fig. 7). Muscle separated the leaflets of the tricuspid and aortic valves in two patients and the leaflets of the tricuspid and pulmonary valves in the other. In two other patients, there was fibrous continuity between the leaflets of the aortic and pulmonary valves in conjunction with either a hypoplastic or an absent outlet septum (Fig. 8).

Criteria for surgical decisions. Earlier studies (7,9) emphasized the importance of how precise identification of the interrelation between the great arteries can be used as a criterion for optimizing surgical technique. In this study, NMR imaging accurately identified this interrelation in 15 patients.

In a more recent study, Sakata et al. (8) used measurements of distances between the attachments of the leaflets of the tricuspid and the aortic and pulmonary valves that, in conjunction with the presence or absence of obstruction within the pulmonary outflow tract, were used as guidelines for determining the type of anatomic repair. In our study, we were able to measure these distances in 13 patients, principally from images in a transverse plane and occasionally from images in the sagittal plane (Fig. 2 to 5 and 7). In the patient with superoinferiorly arranged ventricles and in the patient with an AV septal defect, the distances were not measured. In one other patient, the images were nondiag-



nostic. Although our center does not yet use these criteria or perform the "réparation à l'étage ventriculaire" procedure (16,17), we were able to demonstrate that the surgical outcome predicted from these retrospective measurements correlated well with the actual surgical outcome in those patients who were suitable for and underwent corrective surgery (Table 2).

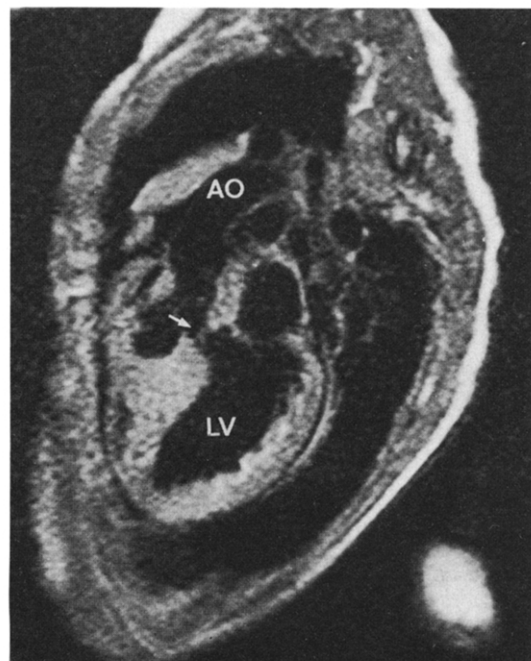
Associated anatomy favoring palliative surgery. Some patients with double-outlet right ventricle and concordant AV connections have additional cardiac abnormalities that increase the risk of death associated with surgical correction. These include the presence of ventricular hypoplasia, superoinferiorly related ventricles and abnormalities of the AV valves (18). Palliative surgery is usually recommended as an initial procedure when these abnormalities are present. Nuclear magnetic resonance imaging was of particular value in identifying these abnormalities.

Cardiac arrangement and ventricular dimensions. In 15 patients, it was possible to determine accurately the morphology, arrangements and relative dimensions of the ventricles. In one patient with an AV septal defect, a large ventricular component of the defect was identified and it was not possible to identify two separate ventricles. Images from transverse sections displaying both ventricles in the long axis were of most value. Additional views of the ventricles in the short axis were sometimes obtained from images from oblique sagittal planes. In 15 patients, the cardiac apex was seen in the left of the thorax with the usual ventricular arrangement. In one patient, the cardiac apex was in the right of the thorax and a section from a standard sagittal plane demonstrated a unique four-chamber view with both ventricles in the long axis, thereby diagnosing a superoinferior ventricular arrangement (Fig. 9). Severe hypoplasia of the morphologic right or left ventricle was not observed.

Abnormalities of the AV valves. Both AV valves were observed in 10 patients. In one patient with a superoinferior arrangement of the ventricles, straddling of the mitral valve

Figure 5. Patient 5. **A**, Oblique sagittal section taken through the plane of the ventricular septum (VS) demonstrating that muscle (single arrow) separates the valvular leaflets of the tricuspid (TV) and aortic (AO) valves. The posterior rim of the ventricular septal defect is also seen (triple arrows). **B**, Anatomic specimen from a patient with similar anatomy (not from this series) displayed in the same orientation seen in **A**. IVS = interventricular septum; VSD = ventricular septal defect.

Figure 6. Patient 14. Ventricular septal defect closing from attachments of tissue from the mitral valve apparatus (arrow). The steep oblique transverse section clearly demonstrates the relation of the ventricular septal defect to the aorta (AO). Left ventricular (LV) hypertrophy is present.



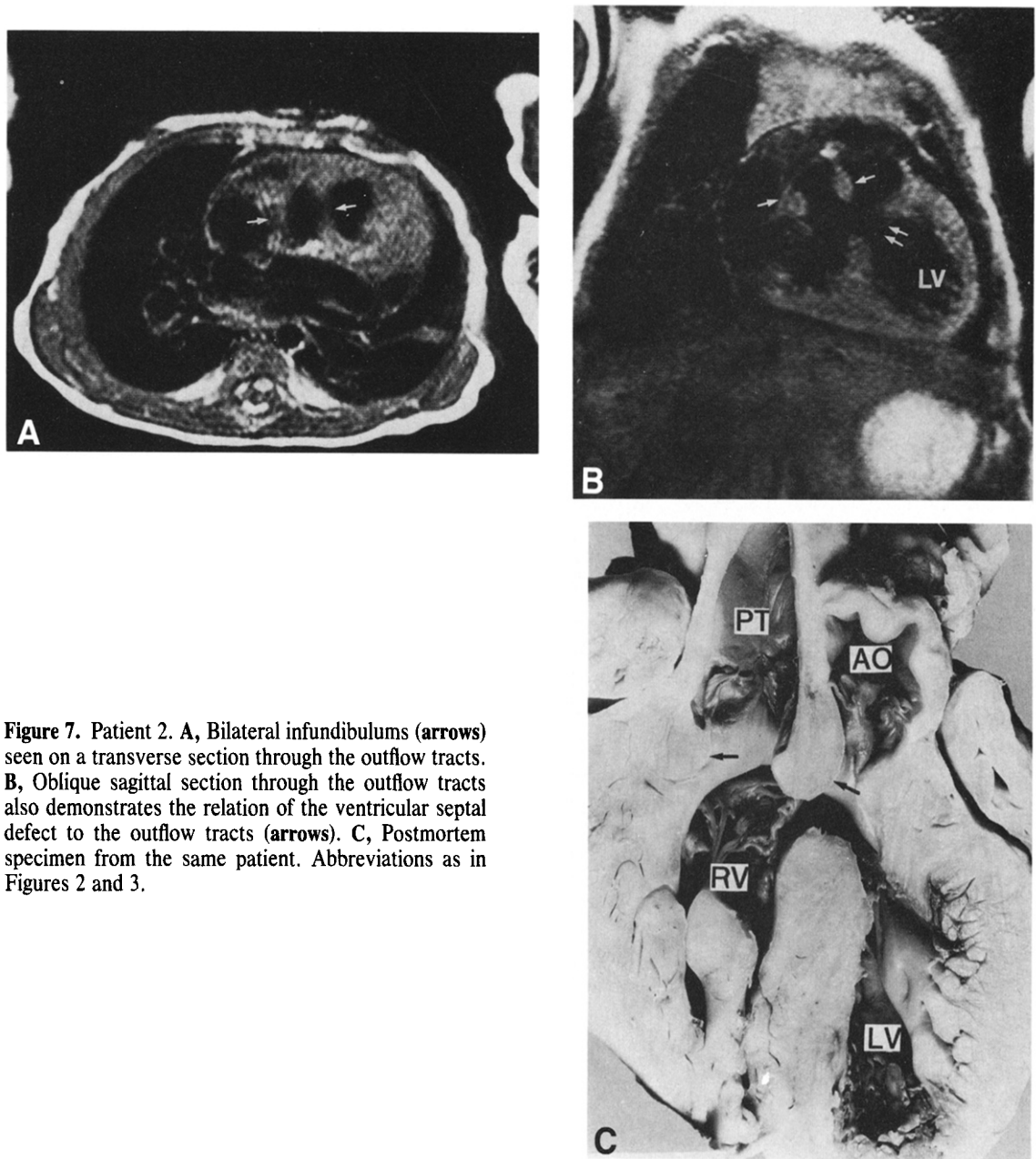


Figure 7. Patient 2. **A**, Bilateral infundibulums (arrows) seen on a transverse section through the outflow tracts. **B**, Oblique sagittal section through the outflow tracts also demonstrates the relation of the ventricular septal defect to the outflow tracts (arrows). **C**, Postmortem specimen from the same patient. Abbreviations as in Figures 2 and 3.

that had not been appreciated with two-dimensional echocardiography was clearly identified from sagittal sections (Fig. 9B). In another patient, with a restrictive ventricular septal defect, the anterior leaflet of the mitral valve was seen to be closely applied to the inferior border of the ventricular septal defect but did not appear to be obviously straddling the septum (Fig. 6). This abnormality was not detected with two-dimensional echocardiography or angiography but was confirmed at surgical correction. No abnormalities were discovered in the remaining eight patients in whom the AV valves were demonstrated. Five of these eight patients have undergone corrective surgery that confirmed the NMR imaging findings. In the six remaining patients, only limited views of the AV valves were obtained. Images from transverse sections were of greatest value in identifying the leaflets of the AV valve and their tendinous cords. These

were demonstrated most clearly from images taken during systole. Other useful images were obtained from oblique coronal sections directed along the plane of the ventricular septum to demonstrate AV valvular leaflets and from sections in an oblique sagittal plane (short-axis view) to show the position of the papillary muscles.

Discussion

During the last 30 years, the overall hospital mortality rate after surgical correction for all patients with double-outlet right ventricle and concordant AV connections has been approximately 30% (18). In recent years, the rate has decreased to approximately 19% (4, 18), which is still disturbingly high. Although one factor that may account for the decrease observed thus far is an improvement in presur-

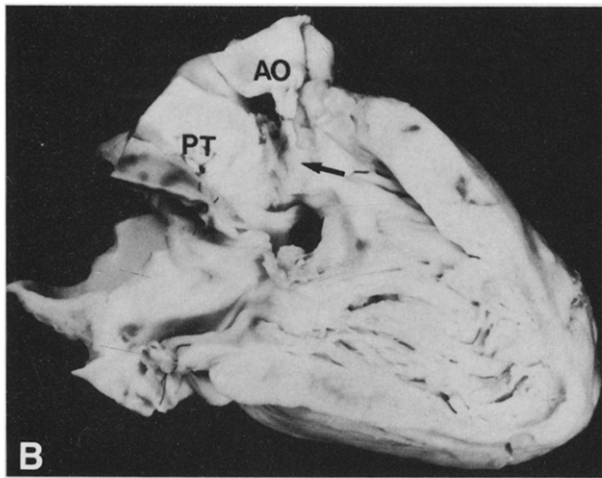
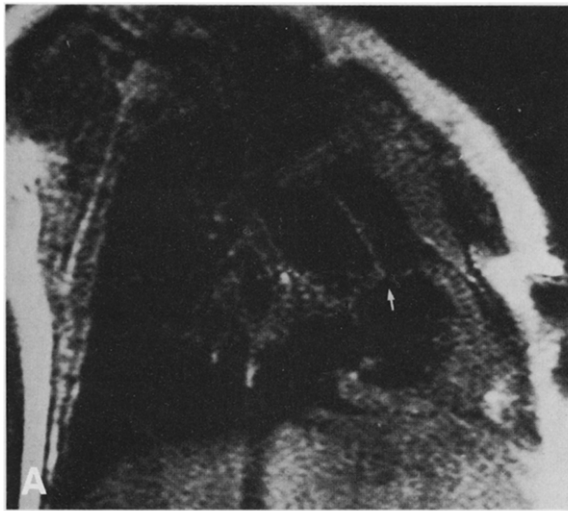


Figure 8. Patient 11. **A**, Oblique sagittal section demonstrating fibrous continuity between the aortic and pulmonary valves (arrow). An infundibulum is not seen. **B**, Postmortem specimen from the same patient. Abbreviations as in Figure 3.

gical diagnosis (18), it is still likely that imprecise presurgical assessment results in inappropriate surgical management in a proportion of cases. Presurgical assessment depends on an accurate demonstration and understanding of the cardiac anatomy; however, the imaging techniques currently used to study these patients have important limitations.

Limitations of Conventional Imaging Techniques

Two-dimensional echocardiography. This is the principal imaging technique used to study this group of patients. With it, Macartney et al. (10) easily demonstrated features that they regarded as of key surgical importance and observed that images from a subcostal window provided the most valuable information. They also described some important limitations of this technique. The first, which applies to

Table 2. Comparison of Predicted Surgical Technique in 13 Patients Based on Criteria Described by Sakata et al. (8) and Actual Outcome

Pt No.	Predicted Surgery	Actual Outcome
1	REV	Palliative BTS
2	IVR	Died before surgery
3	IVR	IVR
4	Arterial switch	Arterial switch
5	REV	Arterial switch
6	IVR	IVR
7	IVR	IVR
8	IVR	IVR
10	IVR	Palliative PA banding
11	Arterial switch	Arterial switch
14	IVR	IVR
15	Arterial switch	No operation, PVD
16	Arterial switch	Palliative PA banding

BTS = Blalock-Taussig shunt; IVR = intraventricular repair; PA = pulmonary artery; Pt = patient; PVD = pulmonary vascular disease; REV = réparation à l'étage ventriculaire procedure.

echocardiography in general, is that echocardiographic windows, particularly the subcostal window, provide limited views in some patients, especially older children, obese patients or patients imaged after thoracotomy. Second, from the subcostal window a great artery may appear to originate from either ventricle in response to slight changes in transducer angle, thereby making diagnosis unreliable in some forms of double-outlet right ventricle. They commented that to determine the true origin of the artery the orifice of the overriding valve must be projected onto the crest of the ventricular septum and this approach requires a projection rather than a cross-sectional technique. Therefore, they used images from parasternal long-axis or precordial short-axis scans to make the diagnosis. Although this method was more reproducible, it led to potential difficulties in distinguishing double-outlet right ventricle from discordant or concordant VA connections. Hagler et al. (11) made the same observation.

Cineangiography. Cineangiography is an invasive technique that requires images to be obtained in several projections to infer the spatial relations among structures of interest. Even then, problems with overlapping structures may lead to difficulties in interpretation. In addition, the tension apparatus of the AV valve is not adequately demonstrated (12,13).

Nuclear Magnetic Resonance Imaging

Previous experience. Previous studies (14,15,19-22) clearly demonstrated the ability of NMR imaging to obtain accurate, high resolution, cross-sectional images of cardiac anatomy in children and infants with congenital heart disease. It is a noninvasive technique that can be used to study patients of all ages. It is not influenced by the depth of structures within the thoracic cavity, deformities of the

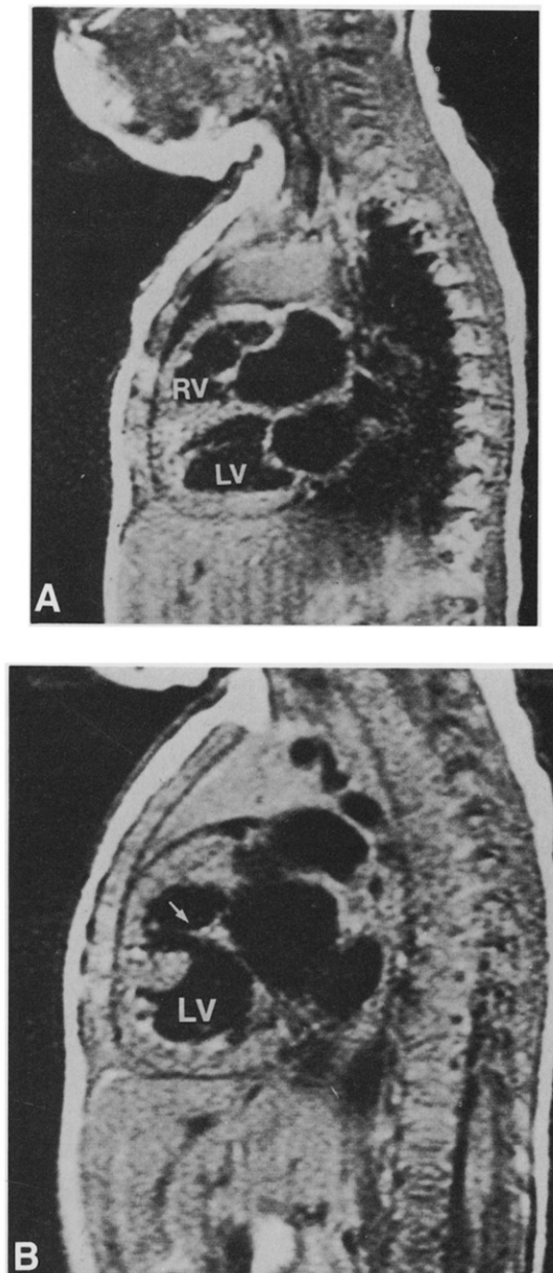


Figure 9. Patient 12. Sagittal sections from a patient with a superoinferior arrangement of the ventricles. **A**, Typical four chamber view normally seen from a transverse section. **B**, The mitral valve (arrow) can be seen overriding the ventricular septal defect. Abbreviations as in Figure 2.

thoracic wall or obesity. It is primarily a cross-sectional imaging technique that is capable of demonstrating anatomy in any plane. By obtaining a series of simultaneous image sections in one plane, however, it becomes possible to appreciate the spatial relations of structures in three-dimensional terms and it is already possible to reconstruct three-dimensional images (23). Despite such advances, the exact clinical role of NMR imaging in the management of patients with congenital heart disease remains under evaluation. Our experience suggests that it can be of important

practical value in the evaluation and treatment of patients with double-outlet right ventricle.

Diagnostic value. An example of the ability of NMR imaging to display the spatial relations of structures is evident when making the diagnosis of double-outlet right ventricle. It is straightforward and reproducible to project images from a transverse plane that demonstrate the great arteries onto images from lower sections in the same plane that demonstrate the site of the ventricular septum, thereby determining the ventricular origin of each great artery.

Importance in preoperative evaluation. Anderson et al. (9) demonstrated that the interrelations between great arteries (which fall into three main subgroups) have a direct bearing on the relation between the ventricular septal defect and the great arteries, which is a major determinant of surgical technique. Yacoub and Radley-Smith (7) also demonstrated that in patients with double-outlet right ventricle whose ventricular septal defect was related to the pulmonary trunk (Taussig-Bing anomaly), the relative positions of the great arteries could be used as a criterion for determining the optimal surgical procedure (either an anatomic switch procedure or intraventricular repair). We have shown that the NMR images were extremely accurate in identifying the relations of the great arterial trunks to each other and to the ventricular septal defect and in demonstrating the size and orientation of the outlet septum when present.

Sakata et al. (8) found that the most important anatomic features in determining optimal surgical technique were the distances between the tricuspid valve and the arterial valves. Because they could not show a fixed relation between these distances and the relative positions of the great arteries, they concluded that it might not always be reliable to use the positions of the great arteries as a criterion for making surgical decisions. They also commented that angiography tended to overestimate these distances and echocardiography was more reliable only when performed specifically to search for these distances. In contrast, we found that it was reliable to measure these distances retrospectively from the NMR images. In all our cases except when the "réparation à l'étage ventriculaire" procedure (which is not performed at our center) would have been recommended, there was a good correlation between the predicted and the actual surgical outcome.

Nuclear magnetic resonance imaging was also capable of demonstrating anatomic features that are associated with an increased risk of corrective surgery (namely, ventricular hypoplasia, a superoinferior arrangement of the ventricles and abnormalities of the AV valve). A particular strength of NMR imaging is the ability to demonstrate the relation of cardiac structures within the context of the thoracic cavity and in particular to demonstrate precise ventricular arrangements. This was illustrated exquisitely in the patient with a superoinferior arrangement of the ventricles. Although none of the patients in our series had ventricular hypoplasia, in a previous study (22) we showed that NMR imaging was better than echocardiography or angiography in demonstrating

ventricular disproportion in patients with an AV septal defect.

In two patients, abnormalities of the mitral valve that had not been detected by two-dimensional echocardiography were identified with NMR imaging. However, in six patients, the AV valve anatomy was not adequately defined. Previous NMR imaging studies (21) have also commented on difficulties in imaging the AV valve. Further experience in this respect is clearly required.

Potential limitation of NMR imaging. NMR imaging may require longer scanning times in studies of patients with double-outlet right ventricle or other conditions characterized by complex cardiac anatomy because many imaging planes may be needed to demonstrate the relevant anatomy. In such situations, adequate sedation must be given to avoid motion artifact that may result in premature termination of the examination. Throughout the study, our sedation regimen was chloral hydrate, initially 50 mg/kg, subsequently increasing to 75 mg/kg. This proved to be safe and was effective in all but one patient.

Conclusions. It is crucial to the management of patients with double-outlet right ventricle that an accurate and precise understanding of the cardiac anatomy is obtained. It is extremely unlikely that two-dimensional echocardiography will be replaced as the first line imaging technique of choice for studying this group of patients. However, either through limitations of this technique or because of the extremely variable and complex nature of the cardiac anatomy, there will always be cases where the echocardiographic images are unclear or uncertainty exists. Nuclear magnetic resonance imaging may well have an important clinical role in these circumstances by providing supplementary information. As a noninvasive technique, it may be preferred to angiography. Further clinical experience in this respect is now required.

We believe that by providing specific, accurate anatomic detail, NMR imaging will contribute greatly to the improved management of patients with double-outlet right ventricle.

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