The spectrum of atrioventricular discordance

A clinical study

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SUMMARY The clinical, investigative, and surgical findings were reviewed in 47 patients with atrioventricular discordance who presented to the Brompton Hospital between January 1962 and June 1981. Although the unifying feature was the atria connecting to morphologically inappropriate ventricles, the hearts differed widely in other respects. In most cases there was the usual visceral and atrial arrangement, but six had a mirror image arrangement of the atria and viscera. Among those patients with usual atrial arrangement (solitus) the aorta was not always anterior and left sided, and ventricular "inversion" was not invariable. Only 35 of the 47 patients also had a discordant ventriculoarterial connexion, the majority of the remainder having a double outlet right ventricle.

The specific diagnosis of atrioventricular discordance depended on echocardiographic and angiographic examination. The other investigations did not distinguish between different ventriculoarterial connexions and were not specific even for the presence of atrioventricular discordance. For those patients with the usual atrial arrangement the anticipated right heart border was present in only just over one third, and the reversal of Q wave progression in the precordial leads of the standard electrocardiogram was found in under a half.

Many patients with atrioventricular discordance progressed normally to adult life. In 20 cases no surgery was performed. The results of total correction showed a mortality of 25% (three cases), including two deaths after a modified Fontan procedure for exceedingly complex associated lesions. The results of surgery in the survivors were excellent, and awareness of the disposition of the atrioventricular conduction tissue made it possible to avoid atrioventricular dissociation.

Morphologists and clinicians alike have been fascinated by congenitally corrected transposition since its first description by von Rokitsky in 1875.¹ Confusion abounds concerning the anomaly, partly because it has been called by many names—such as ventricular inversion² or l-transposition³—and partly because it resembles and has been categorised together with other malformations such as hearts with double inlet to a left ventricle having a left sided rudimentary right ventricle.³⁻⁵ The essence of the lesion is atrioventricular discordance, associated in corrected transposition with ventriculoarterial discordance. But it is well recognised that other ventriculoarterial connexions may be found with atrioventricular discordance and

that the anomalies then produced are "close cousins" of corrected transposition.⁶ In this study we examined the records of all the patients known to have atrioventricular discordance seen at the Brompton Hospital and analysed their clinical and investigative findings together with their natural history, as modified in some cases by surgery.

Patients and methods

Between January 1962 and June 1981, 47 patients with atrioventricular discordance were seen at this hospital. These cases were identified by reviewing reports of all clinical, echocardiographic, catheterisation, surgical findings, and follow up data. In four cases necropsy was also performed. We excluded cases with an overriding left valve when more than half of the valve was connected to the same ventricle as the other atrioventricular valve, because from the

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standpoint of the atrioventricular connexion these cases have double inlet to a left ventricle with a left sided rudimentary right ventricle.

PRESENTING FEATURES

The age of the patients (29 (63%) male, 18 (37%) female) at the first examination ranged from 1 day to 31 years (mean 9 years). The dominant clinical features at presentation are shown in Table 1. In general, patients presented with symptoms when there was severe pulmonary stenosis or atresia with or without a ventricular septal defect or when there was a large ventricular septal defect. Even in these groups, however, some patients did not present until the second decade. As expected the symptoms were non-specific and frequently reflected associated anomalies rather than atrioventricular discordance. The exceptions were those presenting with complete atrioventricular block or left atrioventricular valve regurgitation. One patient who presented with severe cyanosis had ventriculoarterial concordance with atrioventricular discordance and no other associated lesions. In this infant the haemodynamic effects produced by the chamber connexions were comparable to those of complete transposition. One patient, aged 24 years, presented with precordial pain typical of angina pectoris. It was only during coronary arteriography that atrioventricular and ventriculoarterial discordance were diagnosed together with mild pulmonary stenosis. Eight patients, who were completely asymptomatic, were referred because of a cardiac murmur. The diagnosis in all patients was confirmed at cardiac catheterisation, at which complete segmental analysis was achieved.

ANGIOCARDIOGRAPHIC FINDINGS

Forty one patients had usual atrial arrangement (solitus) and six had mirror image atrial chambers (inversus). Of those with usually arranged atria, 28 had left sided hearts, 10 had right sided hearts, and in three the heart was in the mid-line. Of the six patients with

Table 2 Cardiac position, connexions, and spatial relations

	Atrial arrangement	
	Usual (n=41)	Mirror image (n=6)
Cardiac position		
Left chest	28	1
Right chest	10	5
Middle-line	3	—
Aortic arch position		
Left	41	1
Right	—	5
Ventriculoarterial connexions		
Concordant	—	1
Discordant	32	3
Double outlet right ventricle	7	2
Single outlet pulmonary atresia	2	—
Ventricular relations (RV vs LV)		
Left, side by side	18	—
Left, side by side and superior	7	—
Left, anterior	12	—
Left, anterior and superior	2	—
Left, superior	1	—
Right, superior and posterior	—	2
Right, side by side	—	3
Right, anterior and superior	1	—
Right, posterior	—	1
Arterial valve relations (aortic vs pulmonary)		
Left, anterior	28	—
Left, side by side	10	—
Right, anterior	3	4
Right, side by side	—	1
Left, posterior	—	1

mirror image atria, only one had a left sided heart (Table 2). The aortic arch was to the left in all patients with the usual arrangement and to the right in all patients with mirror image atria except one.

The ventriculoarterial connexions are also shown in Table 2. In the patients with usually arranged atria, 32 had ventriculoarterial discordance, seven a double outlet right ventricle, and two a single outlet via an aortic trunk with pulmonary atresia (Fig. 1). Of the six patients with mirror image atria, one had ventriculoarterial concordance, three ventriculoarterial discordance, and two a double outlet right ventricle.

Spatial relations between the ventricles were variable (Table 2). In patients with usual atrial arrangement the morphologically right ventricle tended to be to the left of the morphologically left ventricle, whereas in hearts with mirror image atria the right ventricle was to the right of the left ventricle. But in addition the ventricles were found to be in side by side, superior-inferior, or anterior-posterior relations. One case with a usual atrial arrangement showed crisscross atrioventricular relations,^{7,8} the morphologically right ventricle being to the right and anterosuperior to the morphologically left ventricle.

The relations between the arterial valves were again variable (Table 2). With the usual atrial arrangement the aortic valve was anterior and to the left in 28 cases, left and side by side in 10, and anterior and to the right of the pulmonary valve in three. With a mirror

Table 1 Clinical data of 47 patients with atrioventricular discordance

Symptoms	No of patients	Complicating anomaly	Age
Cyanosis	23	PA* or PS with or without VSD†	1 d–16 y
Dyspnoea	14	VSD with or without TI	2 m–15 y
Murmur (asymptomatic)	8	VSD or PS or TI	2 y–31 y
Syncope	1	Heart block	25 y
Angina	1	PS	24 y

*two patients.

†includes one patient with ventriculoarterial concordance.

PA, pulmonary atresia; PS, pulmonary stenosis; VSD, ventricular septal defect; TI, tricuspid incompetence.

Fig. 1 Different types of ventriculoarterial connexion which may be found with atrioventricular discordance: (a) double outlet right ventricle; (b) ventriculoarterial concordance; (c) ventriculoarterial discordance (congenitally corrected transposition); (d) solitary aorta with pulmonary atresia; and (e) double outlet left ventricle. (c) shows the commonest connexion, a discordant one. All the others except double outlet left ventricle were found in our series. MRA (MLA), morphologically right (left) atrium; PT, pulmonary trunk; MRV (MLV), morphologically right (left) ventricle; AO, aorta; Conf, confluence.

image arrangement of the atria the aortic valve was to the right and anterior in four, to the right and side by side in one, and to the left and posterior in one.

There were no major intracardiac lesions in one patient who had mirror image atria and complete heart block and in the patient with mirror image atria and ventriculoarterial concordance. Both patients, however, had anomalies of the systemic veins, the inferior caval vein being interrupted and continued via the hemiazygos system to the left superior caval vein, the right superior vena cava being absent.

Table 3 Associated lesions. (Figures are numbers of cases)

Ventricular septal defect	
Subpulmonary (29 cases)	
Inlet perimembranous + aneurysm memb. septum	1
Inlet perimembranous	28
Subaortic	2
Non-committed (3 cases)	
Inlet muscular	2
Trabecular muscular (multiple)	1
Combined	2
Total	36
Pulmonary stenosis (24 also associated with VSD)	
Valvar and subvalvar	11 (10)
Valvar	9 (7)
Subvalvar	4 (2)
Aneurysm of membranous septum	4 (4)
Supravalvar, subvalvar, and valvar	1 (1)
Total	29
Other lesions	
Left AV valve regurgitation (3 with Ebstein's)	7
Left AV valve stenosis	2
Straddling left AV valve	5
Straddling right AV valve	1
Common atrioventricular orifice	1
No significant lesion	2
Miscellaneous*	20
Total	38

*Atrial septal defect, 9; persistent ductus arteriosus, 2; bilateral superior vena cava, 4; hemiazygos continuation of inferior vena cava, 4; hypertrophic cardiomyopathy, 1. AV, atrioventricular.

Neither of these had atrial isomerism.

The most common associated lesion was a ventricular septal defect present in 36 (78%) cases. Of these defects, 29 were perimembranous and extended into the inlet septum below the pulmonary valve, two defects were in the outlet septum and immediately subaortic, and three were muscular and localised in the inlet septum not directly related to the great arteries (non-committed). In one case there were multiple trabecular muscular ventricular septal defects, whereas in the final case a muscular defect coexisted with a perimembranous defect (Table 3).

Pulmonary stenosis was the second most common lesion found in 29 (63%) cases. The diagnosis was made when a gradient of more than 30 mm Hg was found across the pulmonary valve or subpulmonary infundibulum. The precise site of stenosis was shown angiographically. In four cases an aneurysm of the membranous septum was the cause of the obstruction to the outlet of the morphologically left ventricle. A ventricular septal defect was present in 24 of the 29 cases with pulmonary stenosis, and in only one was pulmonary stenosis an isolated lesion.

Other commonly associated lesions were atrioventricular valve regurgitation in seven patients—three having severely dysplastic tricuspid valves as seen in Ebstein's malformation—and an overriding and straddling left (tricuspid) atrioventricular valve in five; the other associated lesions are shown in Table 3.

Fig. 2 Long axis echocardiograms (a) during systole and (b) during diastole at right angles to the ventricular septum (four chamber cuts) showing the off setting of the atrioventricular valve attachments (arrowed) when the ventricular septum is intact. Note that in atrioventricular discordance the atrioventricular septum thus formed is between the left atrium (LA) and the morphologically left ventricle (MLV). RA, right atrium; MRV, morphologically right ventricle.

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiographic findings at the first examination showed complete heart block in four patients, atrial rhythm in one, and a PR interval longer than 0.20 in three (Table 4). When the clinical follow up of these latter patients is reviewed no further prolongation of the PR interval was found. Twenty patients had left axis deviation (QRS less than -20°). The diagnosis of ventricular hypertrophy was made according to the age of the patient: 28 had right ventricular hypertrophy (60%), seven left ventricular hypertrophy (15%), and three biventricular hypertrophy. Nine patients had a completely normal electrocardiogram. Left atrial hypertrophy was found in all patients with left atrioventricular valve regurgitation. Q waves were found in V4R and V1 in 23 patients (49% of the cases). The remaining patients had Q waves in V5 and V6 or RS waves all over the precordium (Table 4).

CHEST RADIOGRAPHY

The plain frontal chest radiograph showed variations in cardiac size and contour and in the pulmonary vascular markings according to the presence of associated lesions (Table 4). Forty five per cent of the patients had normal x ray films. The typical straight upper left heart border described in cases of "corrected transposition" or notched left heart border was found according to the connexion and relation of the great arteries (Table 4).

ECHOCARDIOGRAPHY

An essential prerequisite to the diagnosis of atrioventricular discordance by echocardiography was the determination of atrial situs. This was most conveniently achieved by establishing bronchial situs from the chest radiography or by analysing the position of the abdominal great vessels at the level of the tenth thoracic vertebra.⁹ In practice it was assumed that

Fig. 3 Long axis echocardiograms (a) during systole and (b) during diastole at right angles to the ventricular septum (four chamber cuts) in a patient with a perimembranous ventricular septal defect (VSD). Note that the atrioventricular valves are attached to the central fibrous body (CFB) at the same level so that there is no atrioventricular septum. RA, right atrium; LA, left atrium; MRV, morphologically right ventricle; MLV, morphologically left ventricle.

when the pulmonary veins were seen draining normally to one atrium, then this was the morphologically left atrium.

In those patients with an intact ventricular septum apical or subcostal four chamber "cuts" showed a reversed pattern of the differential septal attachments of the mitral and tricuspid valves (Fig. 2). In the normal heart—that is, with atrioventricular concordance—the valve draining the morphologically right atrium has a septal attachment more toward the ventricular apex than that draining the left atrium. In the patients we studied with atrioventricular discordance and intact ventricular septum, the valve guarding the junction of the morphologically right atrium with the ventricles had a higher insertion. With the usual atrial arrangement this high valve attachment was right sided. In those with mirror image atria, however, it was the left sided valve which had a high septal attachment.

It was not possible to find this differential

septal attachment in the cases with perimembranous inlet ventricular septal defects (Fig. 3). In these hearts the septal leaflets of the mitral and tricuspid valves were in fibrous continuity through the central fibrous body in the roof of the defect. In these cases, therefore, the existence of atrioventricular discordance could not be diagnosed from the valve septal attachments. Other criteria did still permit the unequivocal echocardiographic diagnosis of mitral and tricuspid valves, and hence morphologically left and right ventricles. In all the hearts examined the chordae of the mitral valve inserted into two discrete papillary muscles, neither of which was attached to the ventricular septum. In contrast chordae from the tricuspid valve always inserted into the right ventricular aspect of the ventricular septum. These findings permitted ventricular identification even in the patients with perimembranous inlet ventricular septal defects, in which some chordae were in all cases attached to the crest of the ventricular septum. Simi-

Table 4 *Electrocardiographic and radiographic findings*

	Atrial arrangement	
	Usual (n=41)	Mirror image (n=6)
<i>Electrocardiographic findings</i>		
Rhythm		
Sinus	37	5
Complete heart block	3	1
Left atrial rhythm	1	—
PR interval (>0.2 s)	3	—
Axis		
+150° to +90°	18	3
+90° to -20°	8	1
-20° to -90°	10	—
-90° to -150°	5	2
Q waves*		
Positive V4R, negative V6	22	1
Negative V4R, positive V6	7	1
Negative V4R, negative V6	12	4
Ventricular hypertrophy		
Right	25	3
Left	6	1
Both	3	—
None	7	2
<i>Radiographic findings</i>		
Pulmonary vascular markings		
Increased	12	3
Reduced	9	—
Normal	20	3
Cardiac size		
Enlarged	22	3
Left heart border†		
Straight	13	5
Knobbed	12	1
Notched	6	—

*For mirror image atrial arrangement leads V6R and V4.

†Right heart border in mirror image atrial arrangement.

larly, in the five hearts with straddling tricuspid valves chordae inserted into the morphologically right ventricular aspect of the septum. In most hearts a short axis cut from apical or parasternal positions

allowed the delineation of three cusps in the tricuspid valve or only two cusps in the mitral valve. Ventricular morphology was also recognised in some of the hearts from the smooth trabecular pattern of the morphologically left ventricle and the coarse trabeculations of the morphologically right ventricle. The presence of the septomarginal trabecula (moderator band) in the morphologically right ventricle was particularly helpful in this respect (Fig. 4).

Cross sectional echocardiography permitted the identification of ventricular septal defects in all the patients having this lesions who were studied with this technique. It similarly permitted the diagnosis and distinction of the types of pulmonary stenosis.

FOLLOW UP

Non-operated patients

In 20 patients followed up on average for nine years surgery was not performed for various reasons. Three were recently lost to follow up but were asymptomatic when last seen. Of the other 17, six were asymptomatic while three were slightly symptomatic and may require operation in the future. In two patients surgery was contraindicated because of pulmonary vascular disease (pulmonary resistance > 8 units in 100% oxygen), both having multiple ventricular septal defects.

Three of four patients who presented with complete heart block at the first examination did not require pacemaker insertion. The fourth patient, referred initially with symptoms of slight dizziness, required insertion of a pacemaker after five years follow up because her symptoms worsened. One 8 year old girl in this group with congenital heart block had a small

Fig. 4 Long axis echocardiogram at right angles to the septum showing the ventriculoarterial junction (there is a discordant connexion) and demonstrating the prominent moderator band. Ao, aorta; PT, pulmonary trunk; MLV, morphologically left ventricle; MRV, morphologically right ventricle.

ventricular septal defect and aneurysm of the membranous septum and developed bacterial endocarditis after seven years of follow up but responded well to treatment and completely recovered. The other two patients with complete heart block, both having additional left atrioventricular valve regurgitation, were progressing to date without requiring either a pacemaker or surgical correction.

There were two deaths during the period of clinical follow up. A 7 year old girl died suddenly three years after diagnosis. Review of her notes showed that she had a period of intermittent heart block during cardiac catheterisation, which may or may not be relevant. The other death was in a 6 month old girl who had a ventricular septal defect and severe hypertrophic cardiomyopathy. She had not responded to treatment with propranolol.

PALLIATIVE SURGERY

Sixteen patients underwent palliative surgery at a mean age of 18 months (Table 5). Five patients required banding of the pulmonary trunk for cardiac failure and pulmonary hypertension in the first year of life and are all well after a mean period of six years. Only one has so far had subsequent correction. Nine patients required a shunt as an emergency procedure for severe cyanosis or small pulmonary arteries. To date seven were progressing well with no hypoxic events.

Over a mean follow up period of five years one has so far had corrective surgery with a good result. Another patient with pulmonary atresia, however, who had a Waterston shunt, died suddenly three years

after apparently satisfactory palliation. Pulmonary valvotomy was performed in an 8 year old boy presenting with ventricular septal defect and severe valvar pulmonary stenosis. He developed atrioventricular dissociation and subsequently complete heart block after the operation, but his cyanosis was relieved and his exercise tolerance improved. He did not require a pacemaker in four years of follow up.

A 1 day old baby with severe pulmonary stenosis and ventricular septal defect had both a shunt and a pulmonary valvotomy and died during the procedure from severe hypoxaemia.

CORRECTIVE SURGERY

Twelve patients underwent corrective surgery, two after previous palliative procedures (Table 5). Five had an uneventful postoperative recovery and were well after a mean follow up of four years. Three patients developed severe rhythm disturbances in the immediate postoperative period. Of these, one had atrioventricular dissociation requiring insertion of a pacemaker. The second, a 9 year old boy, had had previous banding of the pulmonary artery at 2 months of age. During the immediate postoperative period following complete repair several episodes of fascicular tachycardia occurred, but recovery was complete. The third patient, an 8 year old girl, had a ventricular septal defect patched and a dysplastic Ebstein's like left atrioventricular valve replaced by an artificial (Björk-Shiley) valve. She developed atrial tachycardia but was to date in sinus rhythm though with episodes of atrial fibrillation.

One patient, a 3 year old boy, had his left atrioven-

Table 5 Clinical data of patients undergoing palliative and corrective surgery

Procedure	No of patients	Age	Complications	Subsequent correction
<i>Palliative surgery</i>				
Banding of pulmonary trunk	5	2 m-1 y (median 6 m)		1
Blalock-Taussig shunt (including 5 Gore-tex)	7	1 w-15 y (median 1 y)		
Waterston shunt	2	1 w, 7 y	1 late death (3 y)	1
Waterston shunt and pulmonary valvotomy	1	1 d	Died during surgery	
Pulmonary valvotomy	1	8 y	Intermittent AV dissociation	
<i>Corrective surgery</i>				
Closure VSD, LV-PA conduit	3	1 y,* 11 y, 15 w	Complete heart block (1 case, died)	
Closure VSD, outflow tract patch	1	18 y		
Closure VSD, replacement LAVV (Björk)	1	8 y	Atrial fibrillation	
LV-PA conduit, excision AMS	1	10 y		
LV-PA conduit, closure ASD	1	16 y		
LAVV replacement (Carpentier-Edwards)	1	1 y	Calcification replaced at 3 y (St Jude prosthesis)	
Modified Fontan procedure	2	4 y, 7 y	Both died	
Debanding of pulmonary trunk, closure VSD	1	9 y	Supraventricular tachycardia	
Modified Mustard	1	10 m		

*After previous Waterston shunt.

VSD, ventricular septal defect; LAVV, left atrioventricular valve; LV, left ventricle; PA, pulmonary artery; AV, atrioventricular.

tricular valve replaced, again for Ebstein's anomaly. Initial replacement was with a Carpentier-Edwards porcine xenograft, but this became severely calcified after less than two years. This valve had to be replaced recently, and this was achieved successfully using a St Jude prosthesis.

Three patients died during the first month after operation. One, who had a xenograft conduit placed between the right atrium and the left pulmonary artery, died on the third postoperative day in severe low output and renal failure. The second patient also had a Hancock xenograft valve in a conduit placed between the right atrium and the left pulmonary artery and died on the twenty sixth postoperative day after apparently progressing well. Necropsy showed acute gastrointestinal haemorrhage as the cause of death. In both of these patients, who had a modified Fontan procedure, necropsy confirmed the presence of a very small left ventricle and severe pulmonary stenosis. In addition, one also had crisscross atrioventricular relations and a straddling mitral valve. The third patient, aged 1 year, died in severe cardiac failure on the tenth postoperative day having had a ventricular septal defect patched and a conduit inserted between the left ventricle and the pulmonary trunk. This patient had had a Waterston shunt a month before but continued to have episodes of cyanosis. At necropsy very small pulmonary arteries were found.

Discussion

The 47 patients in this study had one common feature: their atrial chambers were connected to morphologically inappropriate ventricles. In most other respects they differed widely. Most had a usual viscerotrial arrangement, but six had a mirror image arrangement of their thoracoabdominal viscera along with their atrial chambers. Most had ventriculoarterial discordance (transposition) but an appreciable number (20%) had a double outlet from the morphologically right ventricle, whereas in one patient the association with ventriculoarterial concordance produced the haemodynamic pattern usually associated with complete transposition. The associated anomalies were dominated by the triad of lesions traditionally associated with congenitally corrected transposition (ventricular septal defect, pulmonary stenosis, and left atrioventricular valve malformations), but diverse malformations were present in other patients. None the less, the unifying anatomical hallmark of all the hearts, and the feature which determined such an important factor as the disposition of the atrioventricular conduction tissues¹⁰ was the presence of atrioventricular discordance. Indeed, it is only the atrioventricular discordance which was common to all the patients. This in itself illustrates

the inadvisability of grouping such malformations in terms of "l-transposition" or "ventricular inversion." Those who have advocated l-transposition as a suitable term for this group³ may be surprised to find that all of our patients did not have a left sided anterior aorta, even with usually arranged atria. In patients with a mirror image arrangement of the atria a right sided anterior aorta is the rule but is by no means universal. Similarly, those who advocate ventricular inversion as a suitable synonym³ should note that inverted ventricles are present in most but not all of the patients with a usual atrial arrangement and in none with the mirror image arrangement. These facts show that an anomaly unified by the abnormal connexions of the cardiac segments cannot adequately be described in terms of the relation of these segments. This is not to deny the clinical significance of the relations, but simply that in this context they must be described as subordinate to the connexions.¹¹

In terms of the connexions, it is noteworthy that only 35 of the 47 patients studied also had a discordant ventriculoarterial connexion. It is the combination of discordance at atrioventricular and ventriculoarterial levels which can conveniently be described as congenitally corrected transposition. But in our experience this combination is found in only three quarters of patients with atrioventricular discordance. Despite this there were no clinically significant differences noted in the clinical, radiographic, and electrocardiographic features among the patients with or without transposition. This, then, points to perhaps our most clinically significant, and readily obvious, finding. The specific diagnosis of patients with atrioventricular discordance rests upon the echocardiographic and angiographic examination. The other investigations in no way distinguish between the different ventriculoarterial connexions and, in many cases, are not specific even for the presence of atrioventricular discordance. Thus the mode of presentation reflected the associated anomalies rather than the discordance itself. The anticipated straight heart border (left sided in usual atrial arrangement and right sided in mirror image atrial arrangement) was present in only just over one third of patients. The anticipated reversal of Q wave progression in the precordial leads of the standard electrocardiogram was found in less than half of the patients. These various investigations can, therefore, raise the suspicion—in many cases a very high suspicion—of atrioventricular discordance, but the final diagnosis rests upon the echocardiographic and angiographic findings.

Recent studies have elegantly shown the value of angiography, particularly using axial views, in the diagnosis and analysis of hearts with atrioventricular discordance.^{12 13} In our opinion, however, precise

details of the morphology of the atrioventricular junction itself are better shown by cross sectional echocardiography. This technique can detect not only the atrioventricular discordance but also the nature of the associated malformations and details of the mitral and tricuspid valves. A recent collaborative study showed how the characteristic morphology of the ventricular inlets permits not only precise echocardiographic diagnosis of the discordant atrioventricular connexion but also the siting of the ventricular septal defect and the nature of any pulmonary stenosis or tricuspid valve malformation.¹⁴ The variability detected in these lesions in our clinical study mirrors closely the necropsy findings.¹⁵ Other features, however, such as the relative size of the ventricles, their spatial relations, and details of the ventriculoarterial junction are more easily discerned by angiography. Both the angiocardiographic and echocardiographic techniques readily permit the distinction of hearts with atrioventricular discordance from those with a double inlet left ventricle with a left sided rudimentary right ventricle. No longer is there any justification for the grouping together of these malformations.³⁻⁵ Nowadays the surgeon will be less than well pleased if the patient referred to him with "l-transposition" is found also to have a double inlet to the left ventricle.

Our study shows that considering the complexity of the malformation many patients with atrioventricular discordance may progress normally to adult life. Despite the fact that the circulatory pattern is not totally corrected in the presence of associated malformations, only just over half of our patients required palliative or corrective surgery. The results of surgery were also encouraging. One patient died during palliative surgery, partly as a result of an imprudent pulmonary valvotomy producing heart block in a period before we were aware of the unusual position of the atrioventricular bundle.¹⁰ One patient died suddenly three years after a shunt procedure, and one had intermittent atrioventricular dissociation, also after pulmonary valvotomy. Apart from these, to date the remainder are well, two having undergone subsequent surgical correction. Our results of total correction show a mortality of 25%. Two of these three deaths, however, followed modified Fontan procedures for exceedingly complex associated lesions. In the other patients the results of surgery were excellent. Both our results and experience are in keeping with others' reported surgical experience with atrioventricular discordance.¹⁶⁻¹⁸ Like them, since we have been aware of the disposition of the atrioventricular conduction tissues, atrioventricular dissociation has no longer proved a major problem.

Thus our experience suggests that the double discordance does indeed "correct" most of the problems of transposition, at least in the period of our follow

up. Nevertheless, problems certainly remain. We cannot be sure of the fate of the morphologically right ventricle and tricuspid valve in supporting the systemic circulation. In this respect the unusual combination of atrioventricular discordance and ventriculoarterial concordance becomes the ideal for an atrial redirection procedure. The operation corrects the haemodynamic effects of "complete" transposition and at the same time restores the morphologically left ventricle to a systemic role. But these cases are exceedingly rare. In the rest, it is the morphologically right ventricle which must bear the systemic role. Recent studies by the Toronto group suggest that it is ill suited so to do,¹⁹ but Graham has reported normal values in a further group of 10 patients.²⁰ Our next step will be to evaluate morphologically right ventricular function and response to exercise in our large group of patients.

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