Morphological considerations pertaining to recognition of atrial isomerism

Consequences for sequential chamber localisation

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SUMMARY The atrial morphology and venous connections were assessed "blind" in 51 necropsy specimens from patients with visceral heterotaxy. This was compared with bronchial morphology as established by dissection. Six specimens were found to have both atria and bronchi in situs solitus or inversus, and were rejected. In the remainder, atrial isomerism was diagnosed, though this required minor revision of the atrial assessment in two patients. Thirty-four patients had isomeric right atria and bronchi, while 11 had isomeric left atria and bronchi.

In seven cases, splenic status was unknown, but in seven of the remaining 38 (18.4%) atrial isomerism was not associated with either asplenia or polysplenia. Nevertheless, right isomerism was strongly associated with total anomalous pulmonary venous drainage (as is asplenia) and left isomerism was likewise associated with interruption of the inferior vena cava (as is polysplenia). Bilateral superior venae cavae and hepatic veins, and absence of the coronary sinus, were frequent in both forms of isomerism (as they are in asplenia and polysplenia).

These findings suggest that atrial situs can be defined as solitus inversus, right isomerism, and left isomerism. This determination of atrial situs is quite independent of any other abnormalities of visceral situs. The high incidence of anomalies of both venous return and common atrium resulted in presumed complete mixing of blood at atrial level in all but one patient (97.8%), making the haemodynamic connection between atria and ventricles almost always ambiguous. To describe this anatomical connection as ambiguous when there are two ventricles present is therefore no more than a recognition of anatomical and haemodynamic reality.

Visceral heterotaxy presents two fundamental questions to the cardiac morphologist. First, are the atrial chambers of ambiguous or indeterminate morphology,^{1 2} or are they isomeric? Second, if the atria are isomeric is it possible to avoid the necessity for describing the atrioventricular connection as ambiguous? In theory, it might be feasible to describe the connections as concordant or discordant on the basis of analysis of venous connections and ventricular morphology.

A further practical question for the clinician is how best to predict the presence of atrial isomerism

before cardiac catheterisation. While it is well recognised that bronchial morphology correlates well with atrial morphology³⁻⁷ and that bronchial situs can be objectively diagnosed in life,⁵ isolated exceptions to this rule have been reported.^{4 8 9} The real question is not whether there are any exceptions, but how frequent they are.

To investigate these questions we have studied visceral situs, atrial morphology, and venous connections in 45 patients in whom the diagnosis of visceral heterotaxy was made or confirmed at necropsy.

Subjects and methods

The case material came from the necropsy collections at The Hospital for Sick Children, Great

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Ormond Street, the Brompton Hospital, and Killingbeck Hospital, Leeds. Cases selected for study were those which did not unequivocally have situs solitus or inversus, provided that one or both lungs were attached to the heart. The mediastinum and hila of the specimens were thoroughly dissected by one of us (FJM), so as to expose the pulmonary veins, pulmonary arteries, and bronchi. Bronchial morphology was assessed by noting the course of the central pulmonary artery as it continued, travelling posteriorly and inferiorly, into the lower lobe pulmonary artery. If this arterial pathway passed between two bronchi (upper lobe and middle lobe) the bronchus was designated eparterial. If it crossed above all bronchi (specifically the upper lobe bronchus) the bronchus was designated hyparterial.¹⁰

In this way, 44 specimens were obtained with isomeric bronchi (that is bilateral hyparterial or eparterial bronchi). In a further specimen one bronchus was presumed hyparterial since the lung removed at necropsy was bilobed, and the other bronchus was definitely hyparterial. There were six further specimens with lateralised bronchi, in all of which the presence of splenic abnormalities or anomalies of abdominal situs not corresponding with thoracic situs had raised doubts about whether atrial situs was isomeric or lateralised.

These 51 specimens were all arranged so that only the atrial appendages could be seen and then RHA assessed atrial morphology, "blind" without any other knowledge of the specimen. In all six patients with lateralised bronchi, assessment of atrial morphology from the external appearance of

the appendages alone indicated lateralised situs, which was confirmed on inspection of the internal anatomy of the atria. The atrial situs corresponded to bronchial situs in all six, and was solitus in four and inversus in two. These hearts are not described further.

In the remaining 45 specimens, atrial morphology was assessed from the external appearance of the appendages alone in 39, and the internal morphology was not examined until later. After inspection of the internal morphology in the remaining six, atrial isomerism was diagnosed in all 45 patients (see Results).

In 20 of these specimens the abdominal viscera remained attached to the specimen, making precise identification of abdominal situs and venous drainage below the diaphragm easy. In the remainder, such information was gleaned from the necropsy reports, from descriptions of cardiac catheterisation, and from angiocardiology or venography.

The data were then punched on to cards and analysed through the University of London Computer Centre, using the *Statistical package for the social sciences*¹¹ employing the χ^2 and Fisher's exact test for cross-tabulation.

Results

MORPHOLOGY OF BRONCHI (Fig. 1)

All cases with two lungs had isomeric bronchi. In 34 they were of right morphology and in 10, of left. The remaining case had a left bronchus in one lung and was said to have had a bilobed lung on the other side.

Fig. 1 Photographs illustrating the difference between isomeric bronchi of right morphology (Fig. 1a) and isomeric bronchi of left morphology (Fig. 1b). The bronchi of right morphology (closed star) are short and the pulmonary arteries continuing their lower lobe branches pass between the first branches of the bronchi and the intermediate stem bronchi. The bronchi of left morphology (Fig. 1b) are both long (closed stars) and the pulmonary artery supplying the lower lobes curves over the bronchi before they give their first (upper lobe) branch (open stars).

Fig. 2 Photographs of the atrial appendages from a heart with situs ambiguus and right isomerism. Both appendages are blunt and both join with the atrial chamber along a broad front. Though there is a fold in the left-sided morphologically right appendage (Fig. 2b) it can clearly be seen that this is still a blunt appendage and is of right morphology. SVC, superior vena cava.

In several cases with bilateral eparterial bronchi, the right upper lobe pulmonary artery originated early (as frequently occurs in normal individuals)¹² and gave the erroneous initial impression that the bronchus was hyparterial since it lay below the right upper lobe pulmonary artery. Once dissection had been carried immediately distal to the first bifurcation of the main bronchus, unequivocal diagnosis of the bronchial morphology was straightforward. In two patients, there was a pre-eparterial bronchus on each side (designated as morphologically right).

MORPHOLOGY OF ATRIA (Fig. 2 to 4)

This was determined on the shape of the atrial appendages, and the morphology of the junction of the trabecular part of the appendage with the smooth walled parts of the atria. Venous connections and septal morphology were not considered because the venous connections were frequently bilateral and the coronary sinus usually absent, while the atrial septum was frequently severely reduced in size or absent.

An atrial chamber was considered to have right atrial morphology when its appendage was blunt,

Fig. 3 Photographs showing the morphology of the atrial appendages in a heart with situs ambiguus and left isomerism. Both appendages are narrow and sinuous and join with the atrial chamber along a narrow front. Abbreviations as before.

and had a broad junction with the smooth walled atrium. This type of junction was marked by a prominent muscle band protruding into the cavity of the chamber, the crista terminalis.

Left atrial morphology was considered present when the left atrial appendage was long and thin with constrictions along its length. Such appendages had a more constricted junction with the smooth atrium. Though when such left atrial chambers were dilated, the junction had a pectinate appearance, a prominent crista terminalis was not identified. In all the hearts studied, two atrial chambers were identified, on the basis of there

CORRELATION OF ATRIAL WITH BRONCHIAL MORPHOLOGY

In one of the 39 specimens assessed externally, initial examination of the atrial appendages led to an incorrect diagnosis of situs solitus. This was because the right-sided atrial appendage had been rendered blunt by atriotomy at attempted surgical repair of a complete atrioventricular defect. When the internal anatomy was subsequently reviewed, it was clear that there was left isomerism. In the remaining 38, atrial morphology assessed from external appearance alone corresponded with bronchial morphology.

Fig. 4 The opened right-sided atrial appendages illustrated in Fig. 2a and 3a. The atria of right morphology (Fig. 4a) shows clearly a well-formed crista separating superior vena cava from the atrial appendage. Note that the posterior continuation of the crista has pectinate muscles inserted along its length producing an appearance reminiscent of a comb. This arrangement is not seen in the morphologically left atrium illustrated in Fig. 4b. In addition there is no crista separating the pectinate muscles of the atrial appendage from the superior vena cava in this atrium. The pectinate muscles are anteriorly positioned and fade out as they pass posteriorly. This is in contradistinction to the comb-like appearance illustrated in Fig 4a in the morphologically right atrial chamber.

being two atrial appendages, each with a junction with the smooth-walled portion of the atrium. These atrial chambers were always both of the same morphology, either right morphology (right atrial isomerism) or left morphology (left atrial isomerism). This is not to be construed as indicating the presence of two normal right or left atria, simply that the atrial chambers had bilateral morphological characteristics of right or left type.

In the six patients in whom the external appearances of the atrial appendages were equivocal, the internal anatomy of the atria was viewed with the remainder of the heart still hidden. In one of these hearts, inversus atria were diagnosed on blind assessment in the presence of bilateral hyparterial bronchi. On review, it appeared that a flap of atrial tissue had been mistaken for crista terminalis, so the final diagnosis was left isomerism. In the

remaining five, atrial morphology corresponded unequivocally with bronchial morphology.

In summary, no case was found with isomeric bronchi which did not have isomeric atria.

ATRIAL MORPHOLOGY AND OTHER INDICANTS OF SITUS

(a) Lung lobulation (Table 1)

Though there was a highly significant association between bilobed lung and left atrium and trilobed lung and right atrium, there was a 14.4 per cent failure rate of lung lobulation as a predictor of atrial situs.

Table 1 Atrial anatomy and lung lobulation

	Right atrium	Left atrium
Bilobed lung	7 (30%)	16 (70%)
Trilobed lung	61 (91%)	6 (9%)

(b) Splenic status (Table 2)

Polysplenia was strongly associated with left isomerism, and asplenia with right isomerism ($p < 0.0001$). Nevertheless there were four of 38 cases (10.5%) with a single spleen who had right isomerism, with the spleen hypoplastic in only one. Furthermore, in three out of 10 cases (30%) with left isomerism, there was a single spleen. There were thus seven out of 38 patients (18.4%) in whom atrial isomerism was not associated with either asplenia or polysplenia. One patient with polysplenia had right isomerism.

Table 2 Spleen in atrial isomerism

	Single (R)	Single (L)	Multiple (R)	Multiple (L)	Absent
Right isomerism*	2 (7%)	2 (7%)	1 (4%)	0	23 (82%)
Left isomerism†	1 (10%)	2 (20%)	2 (20%)	5 (50%)	0

* Six splenic state unknown.

† One splenic state unknown.

(c) Hepatic position (Table 3)

This was lateralised in 20 of 37 patients (54.1%) in whom it was recorded.

Table 3 Liver position in atrial isomerism

	Right	Left	Central	Unknown
Right isomerism	7 (26%)	6 (22%)	14 (52%)	7
Left isomerism	4 (40%)	3 (30%)	3 (30%)	1

$\chi^2 = 1.4$ (not significant).

(d) Gastric position

There was no pattern noted for gastric position in the series as a whole. In any individual case, this was often variable because the stomach was suspended on a long dorsal mesogastrium, and could move easily from one side to the other.

(e) Gall bladder

This was absent in one case with left isomerism out of eight in which its morphology had been noted.

SYSTEMIC VENOUS RETURN

An interrupted inferior vena cava (Table 4) occurred in seven out of 11 patients (63.6%) with left isomerism, and in none with right isomerism. The hepatic veins (Table 5) frequently either straddled the atrial septum, or drained separately to both atria or both sides of a common atrium.

Table 4 Azygos continuation in atrial isomerism

	None	Right	Left
Right isomerism	34 (100%)	0	0
Left isomerism	4 (36%)	1 (9%)	6 (55%)

$\chi^2 = 25.6$ ($p < 0.0001$).

The superior venae cavae were bilateral in 20 out of 45 patients (44.4%), there being an atretic cord on one side and a patent vessel on the other in a further three. In only one patient (with left isomerism) did one superior vena cava (the left) drain to a coronary sinus. In all others, the coronary sinus was absent, and the superior venae cavae drained to the "top corners" of the atria in close proximity to the atrial appendage. No patient with right isomerism had a coronary sinus, though two with left isomerism did. The type of isomerism was not associated at a statistically significant level with either absence of the coronary sinus or bilateral superior vena cava.

Table 5 Hepatic venous drainage in atrial isomerism

	Unilateral	Bilateral	Straddling
Right isomerism	25 (73%)	6 (18%)	3 (9%)
Left isomerism	8 (73%)	2 (18%)	1 (9%)

$\chi^2 = 0.003$ (not significant).

PULMONARY VENOUS RETURN

In seven out of 11 patients (63.6%) with left isomerism, two pulmonary veins returned to each atrium or each side of a common atrium, whereas in the remainder all pulmonary veins drained to one atrium. Bearing in mind that there were

Fig. 5 Photographs illustrating the different mode of insertion of the pulmonary veins into the atrial chambers in left isomerism and right isomerism. Fig. 5a shows the left-sided atrial chamber (of left atrial morphology) in a patient with left isomerism. The left superior vena cava is seen entering the roof of the atrium between the appendage and a broad posterior atrial wall. There is azygos continuation for the inferior vena cava to the left-sided cava. The pulmonary veins enter the broad posterior wall of the chamber with a considerable distance between left pulmonary veins and the cava and between left pulmonary veins and right pulmonary veins (not visible in this photograph). Fig. 5b shows the drainage of the pulmonary veins from both lungs (bilat PV) to the roof of the atrial chambers in a patient with situs ambiguus and right isomerism. Note the right-sided crista. The pulmonary veins enter closely together into the roof of the atrial chambers, and are quite differently inserted from either the normal heart or the patient illustrated with left isomerism.

bilateral left atria, pulmonary venous drainage could be regarded as normal, particularly in the way the veins returned to the heart. There was the normal wide area of posterior atrial wall between the pulmonary veins when the heart was viewed from the back (Fig. 5a)

In contrast, pulmonary venous return was anomalous in all but one patient (97.0%) with right isomerism. The site of pulmonary venous return was unknown in one, but was presumably anomalous, since no pulmonary veins were seen entering the atria. In eight patients out of 33 (24.2%), though the veins returned to one or other atrium or a common atrium, the connection with the heart was abnormal in that, viewed from behind, there was a very small area of atrial wall between the points of entry of the pulmonary veins (Fig. 5b). In three of these patients the veins became confluent before entry into the atrial mass via a single, obstructed, common vein. In a further patient half the pulmonary veins returned to the atrial mass and half to the portal vein. The sites of pulmonary venous return in right isomerism are given in Table 6. Table 7 compares pulmonary venous return in the two forms of isomerism.

Table 6 Pulmonary venous drainage in atrial isomerism

Superior vena cava	9
Infradiaphragmatic (6 portal)	8
Atrium	8
Mixed	6
Vertical vein	2
Unknown	33
	1

Pulmonary venous return was judged obstructed on anatomical grounds in 12 out of 31 cases (38.7%) of right isomerism. In three cases it was not known whether there was any obstruction.

Table 7 Anomalous pulmonary venous drainage in atrial isomerism

	None	Partial	Complete	Unknown
Right isomerism	1 (3%)	0	32 (97%)	1
Left isomerism	4 (36%)	7 (64%)	0	0

$\chi^2 = 39.7$ ($p < 0.001$).

OVERALL VENOUS RETURN

We defined a "solitus" venous connection as one in which the systemic veins drained to the right-sided atrium (or right side of a common atrium) and the pulmonary veins drained to the left-sided atrium (or left side of a common atrium). An "inversus" connection was the inverse of the above. Otherwise the venous connection was termed "ambiguous". An "ambiguous" connection was extremely common (Table 8).

ATRIAL SEPTATION

In 14 patients with right isomerism (42.4%), and

in none with left isomerism, there was a simple, narrow, strand of atrial tissue running across the atrial cavity from immediately anterior to a common atrioventricular orifice to the posterior atrial wall a little above the common atrioventricular orifice (Fig. 6). In four further patients with right isomerism, this simple strand became an extended strand by broadening out superiorly and inferiorly at its posterior end. The superior end of the extension ran to a point just medial to the entry of one superior vena cava, and the inferior end ran to a point just medial to the entry of a hepatic vein.

In two patients with right isomerism there was a superior rim of atrial septum only, less than 2 mm in depth.

The degree of atrial septation is shown in Table 9. For the purpose of assessing the haemodynamic state, a common atrium was held to be present when the maximum extent of the atrial septum was a superior rim or extended strand. Thus common atrium was present in 25 (56.8%) cases, 21 with right isomerism and four with left isomerism.

Fig. 6 Photograph of the atrial chambers opened from behind and retracted upwards in a patient with situs ambiguus and right isomerism. The bilateral cristae are seen in the roof of the atrium. There is a common atrioventricular orifice draining both atrial chambers to the ventricular mass. The atrial chamber is crossed by a strand of tissue passing from the posterior atrial wall to the anterior atrioventricular junction. This is the only evidence of a septum in these atrial chambers.

Table 8 Venous connections in atrial isomerism

	"Solitus"	"Inversus"	"Ambiguous"
Right isomerism*	0	1 (3%)	32 (97%)
Left isomerism	2 (18%)	1 (9%)	8 (73%)

* Pulmonary venous return unknown in one.

OVERALL MIXING OF VENOUS RETURN

In an attempt to correlate anatomy with haemodynamics, we presumed that if there was a common atrium or an "ambiguous" venous connection or an absent or imperforate atrioventricular valve, there would be complete mixing of venous return from the body and lungs. Otherwise there would be streaming.

Thus defined, complete mixing was present in 44 out of 45 patients (97.8%). The only patient in whom streaming was present at atrial level also had pulmonary atresia, and therefore complete mixing at ventricular/great artery level.

HEART POSITION

The main bulk of the heart was to the right in 13 out of 45 patients (28.9%), to the left in 31 out of 45 (68.9%), and in the middle in one. There was no significant difference between right and left isomerism in this respect.

AGE AT DEATH

The median ages at death were 1.1 months for right isomerism and 1.75 months for left isomerism.

Discussion

The cardiac lesions associated with visceral heterotaxy are of extreme complexity, so, for the sake of clarity, we have restricted ourselves in this presentation to consideration of visceral situs, venous return to the heart, and atrial morphology. The anatomy from the atrioventricular valves downstream is described elsewhere.¹³

It is now generally agreed^{4 8 14 15} that the method of sequential chamber localisation introduced by Van Praagh *et al.*¹⁶ is the best way of classifying congenital heart disease. This method starts with,

Table 9 Atrial septum in atrial isomerism

	None	Strand	Superior rim only	Primum ASD	PFO or intact
Right isomerism	1 (3%)	18 (55%)	2 (6%)	9 (27%)	3 (9%)
Left isomerism	4 (36%)	0	0	5 (46%)	2 (18%)

ASD, atrial septal defect; PFO, patent foramen ovale.

and is therefore, founded upon, the identification of atrial situs. Yet no agreement exists on how many types of atrial situs exist. While all aver that situs solitus and inversus are discrete entities, some have suggested¹⁷ that these two categories alone may be sufficient. Others have added situs indeterminatus,^{1 15 16} either encompassing both polysplenia and asplenia^{1 15} or else excluding polysplenia.¹⁶ Others maintain that situs ambiguus consists of two separate entities, categorised either as asplenia and polysplenia^{3 4} or as right and left isomerism.^{7 9}

We suggest that the relative merits of these alternatives can be assessed by asking the following four questions, in descending order of importance. Can they be reliably distinguished; are they based upon anatomical facts; are they of more than academic importance; and do they relate to the heart?

The concept of there being only two forms of atrial situs¹⁷ falls at the first fence. This is because the suggested criterion for the morphologically right atrium is the termination of the supradiaphragmatic inferior vena cava.¹⁸ If there are only two types of situs, then it is essential to be able to trace the inferior vena cava to either one or the other atrium. But our series includes four patients in whom the supradiaphragmatic inferior vena cava straddled the atrial septum to connect with both atria. In a further five patients, the inferior vena cava was interrupted and hepatic veins separately entered both atria. There was no way in these cases of deciding which hepatic vein was in reality the supradiaphragmatic inferior vena cava. So all of these patients, according to the criterion of drainage of the vena cava, had right isomerism, which is neither situs solitus nor inversus. There must therefore be at least three types of situs. But are these three types solitus, inversus, and asplenia (right isomerism)?

If the asplenia syndrome is the only form of situs indeterminatus (ambiguus), then all cases of "polysplenia" must have situs solitus or situs inversus. This proposition would not account for the distinctive morphological appearances of the atria in left isomerism^{3 4 9} confirmed by our own studies, nor for the abnormal site and size (or even absence) of the sinus node in this condition.¹⁹ Even if atrial morphology were ignored, it would be impossible on this basis to classify patients with polysplenia who had the abnormalities of inferior vena caval and hepatic venous drainage described above. The very fact that "polysplenia" is neither situs solitus nor inversus nor asplenia means that a further category of situs is necessary. Therefore, is "polysplenia" a variety of situs ambiguus? Since

it forms a syndrome characterised not only by abdominal and thoracic abnormalities, but also by a constellation of cardiac defects, we agree with Stanger *et al.*⁴ that it is indeed a variety of situs ambiguus.

We are then left with the question of how to subdivide and describe those patients who have neither atrial situs solitus nor inversus. The strongest argument in favour of retaining the blanket term "situs indeterminatus" would be that it is an honest recognition of uncertainty. This concept seems to us to have great merit in describing general visceral situs. Our study has confirmed repeated observations by others,^{1 3 4 20-22} that there is no consistent pattern of arrangement of either abdominal viscera or even of lung lobes in situs ambiguus. In short, there is visceral heterotaxia. But our results also show that this uncertainty need not be extended to encompass the bronchi and atria. The importance of bronchial anatomy in visceral heterotaxy was first noted by Brandt and Liebow,¹⁰ and subsequently confirmed in small series of patients by others.²²⁻²⁴ The only large series of personally studied patients with situs ambiguus in whom bronchial anatomy was reported is that by Stanger *et al.*,⁴ who found isomeric bronchi in all of 35 patients with asplenia or polysplenia. We have found bronchial isomerism in all of 45 patients with isomeric atria. Thus, though one case has been described in which the patient had asplenia and isomeric atria but solitus bronchi,⁹ there does appear to be very great general consistency in the bronchial branching pattern in situs ambiguus. Furthermore this takes two distinct forms, one with bilateral right, and one with bilateral left bronchi.

As to the consistency of atrial morphology, there is no question that in individual cases of visceral heterotaxy, the general disturbance of atrial anatomy can make identification of the atria difficult. But by assessing the morphology blind, and including patients with lateralised situs as well as those with atrial isomerism in the assessment, we have shown that an experienced observer can reliably identify atrial isomerism. Since that assessment corresponded exactly to bronchial morphology, and because it was so different for patients with on the one hand, anomalous pulmonary venous return, and on the other hand interruption of the inferior vena cava, we contend that right and left atrial isomerism are biological and anatomical realities and not simply "convenient mnemonics".²

This view is supported by examination of the conduction system.^{19 23 25} Right isomerism is characterised by bilateral sinus nodes, which are right-sided structures, whereas in left isomerism the node

is either absent or else displaced and hypoplastic.

For the reasons outlined we endorse wholeheartedly the view⁴ that situs ambiguus should be divided into two categories. We do not consider, however, that "polysplenia" and "asplenia" are the optimal terms for these sub-categories. It is our preference to divide atrial situs ambiguus into right isomeric and left isomeric forms. There are many reasons for this.

Firstly, we lose no accuracy in predicting other associated cardiac anomalies. All the associations we have shown with right isomerism have previously been described for asplenia^{3 4 23 26 27} while those for left isomerism have been described for polysplenia.^{3 4 22 26} This applies not only to anomalies of venous connections and atrial morphology presently described but also to the anatomy of the atrioventricular valves and the region distal to them.¹³

Secondly, we are describing a segment of the heart, which is surely more appropriate for a cardiac nomenclature than one incorporating a description of the spleen.⁴

Thirdly, the relation between left isomerism and polysplenia, right isomerism and asplenia is close, but not one to one. Indeed we found 18.4 per cent of patients with isomeric atria and bronchi who did not have asplenia or polysplenia. The asplenia syndrome without asplenia (sic!) has also been described by others.^{3 28}

Finally, diagnostic "labels" affect the way in which patients are investigated. Cardiologists are becoming accustomed to being able to attach exceedingly accurate diagnostic labels to their patients. The label "polysplenia" cannot be attached with certainty until the diagnosis of polysplenia is established. Since this is not always possible with splenic scintigraphy because of inadequate resolution,^{5 29 30} the temptation is to proceed to splenic arteriography.³¹ This we regard as unfortunate, because as far as we know, polysplenia per se does not harm the patient and its demonstration will not affect management. By contrast, it is very important to know before cardiac catheterisation whether or not there is left or right isomerism because this immediately enables a number of predictions to be made about the cardiac anatomy. The value of recognition of a strong possibility of total anomalous pulmonary venous return or interruption of the inferior vena cava before cardiac catheterisation has already been stressed.^{5 32}

It has been shown that bronchial visualisation provides a completely objective means of assessing bronchial morphology,⁵ and that this is usually simple even in infants.³² These studies provide the

proof that this in turn reliably predicts atrial morphology. We accept that occasional exceptions to this rule exist,^{4 8 9} but their existence is no reason for carrying out selective atrial appendage injections of contrast medium⁸ in every patient. Rather, these should be performed when there are good grounds for suspecting a failure of prediction, for example, a hypothetical patient with laevocardia, a patent foramen ovale, normally related ventricles, and normally connected great arteries, who has isomeric right bronchi.

RELEVANCE OF ASPLENIA

While we take the view that splenic status is irrelevant to the segmental description of the heart, this is not to suggest that it does not matter to the patient, since asplenia carries with it an increased risk of overwhelming infection, particularly by the pneumococcus, and long-term prophylactic antibiotics may be indicated.³³ In several of the patients known to have asplenia in this study, Howell-Jolly bodies were sought in the peripheral blood and not found, so they are not completely reliable as predictors of asplenia. We suggest therefore that patients with either isomeric right bronchi or persistent Howell-Jolly bodies in the blood smear should be considered for long-term antibiotics on the presumption that they have asplenia or splenic hypoplasia.

EFFECT UPON THE TYPES OF ATRIOVENTRICULAR CONNECTION

Use of the terms "concordant" and "discordant" to describe the atrioventricular connection was first introduced by Van Praagh *et al.*¹⁶ but then abandoned because they were not applicable in situs ambiguus.² We have retained them¹⁴ because they convey anatomical and haemodynamic information. In the majority of cases of atrial isomerism¹³ the type of atrioventricular connection can be very simply described as double inlet, absent left connection, or absent right connection.¹⁴ It is only in those hearts with two ventricles that the problem arises.

As indicated in the introduction, if it could be shown that the venous return in atrial isomerism could be considered lateralised, then the terms concordant and discordant could still justifiably be used. Our results show conclusively that this is not the case. The anatomy in all but one patient (97.8%) was such that complete mixing of pulmonary and systemic venous blood would have occurred at atrial level. For this reason, we see no alternative to describing the type of atrioventricular connection as ambiguous in hearts with isomeric

atria and two ventricles. This term conveys much anatomical information, indicating as it does that two ventricles are present. And no haemodynamic information is lost in the majority of cases, because pulmonary and systemic venous blood have already mixed completely at atrial level. There may be a tiny minority of patients in whom further brief haemodynamic information might be useful.

An example would be a patient with an anomaly resembling in every way a typical ostium primum atrial defect except that there was interruption of the inferior vena cava with right-sided azygos continuation, left isomerism, and polysplenia. To describe this heart as left isomerism-ambiguous-concordant (quasi normal situs and connections) conveys considerably more information than the pure segmental notation, whether this be ambiguous-ambiguous-concordant,¹⁴ polysplenia-d-loop(L)-d-normal,⁴ or (A-D-S).³⁴ This extremely rare and eminently soluble problem seems to us a poor reason for abandoning the concept of concordance and discordance² which we and others³⁵ have found most helpful.

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