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Abnormalities of the spleen in relation to congenital malformations of the heart: a survey of necropsy findings in children

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Abstract

A series of 1042 reports of necropsies on children dying at Children's Hospital of Pittsburgh was reviewed. In each case, note was taken of the status of the spleen, the lobation of the lungs, the arrangement of the bronchi, the morphology of the atrial appendages, and the presence of any congenital malformations of the heart and great vessels and of any malformations of the abdominal organs. There was isomerism of the left atrial appendages in eight (0·77%), 13 (1·25%) showed isomerism of the right appendages, and seven (0·67%) had multiple spleens without having isomerism of the atrial appendages. Unexpectedly, a normal spleen was found in one patient with isomerism of the right appendages and also in a patient with isomerism of the left appendages. In one patient with isomeric left atrial appendages there was no spleen. The review showed that the morphology of the atrial appendages, and hence the arrangement of the atria, is not accurately predicted by the type of spleen. The arrangement of the atrial appendages is the most reliable guide to the recognised combinations of congenital cardiac malformations previously described as "splenic syndromes". Because there is no certain way of predicting all the malformations in patients with complex congenital heart disease, it is advisable to record separately for each patient the details of lobation of the lungs, the bronchial and atrial arrangement, anomalies of the heart and great vessels, the type of spleen, and any abnormal arrangement of the abdominal organs.

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It is now well recognised that complex congenital malformations of the heart are associated with abnormalities of the spleen and abnormal arrangement of the other thoracoabdominal organs.[1–3] Although it became fashionable to call these cardiac syndromes "asplenia" and "polysplenia",[4] it has since become clear that these are imperfect terms to describe the heart and its contained anomalies. This is because some patients without spleens have the constellation of lesions expected in "polysplenia", whereas the malformations expected in "asplenia" can be found in patients with single or multiple spleens.[5 6] The presence of right or left isomerism of the atrial appendages[6–8] is a much more accurate guide to the characteristic syndromes. Furthermore, some patients with multiple spleens and left isomer-

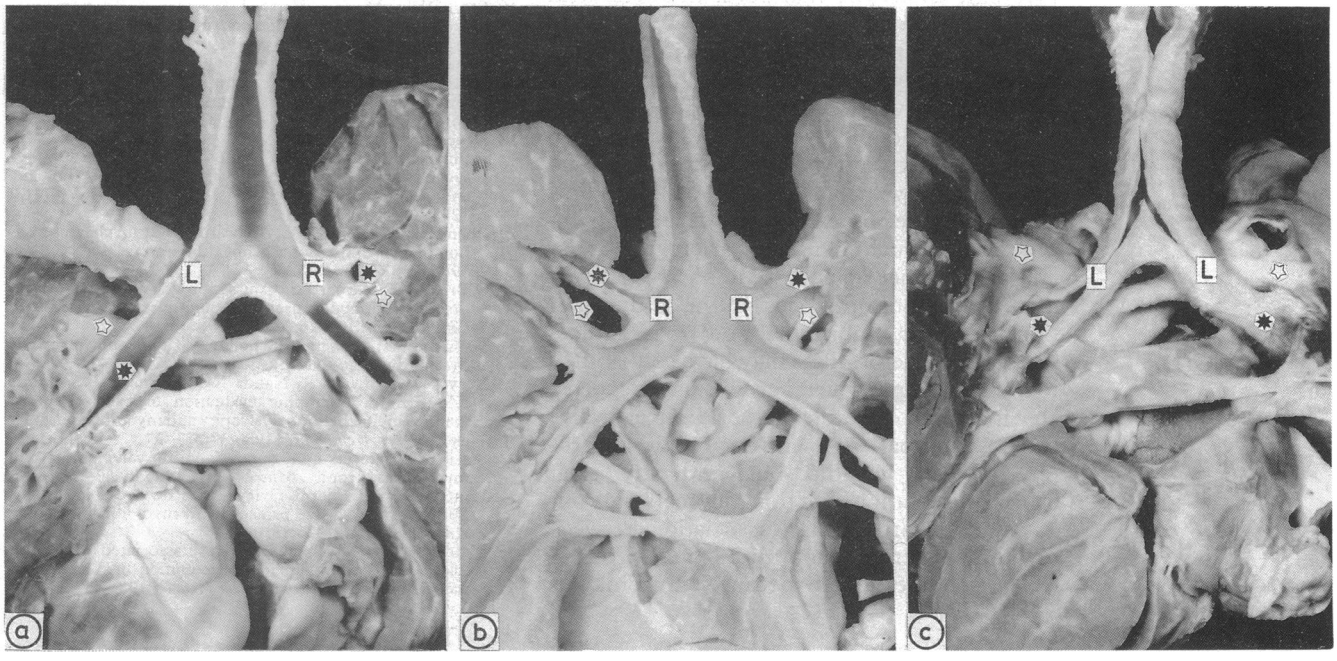


Figure 1 The arrangements of the bronchi are shown from behind in (a) a normal heart, (b) a heart with isomerism of the right bronchi, and (c) a heart with left bronchial isomerism. The left bronchus (L) is longer than the right (R). The morphologically left bronchus is below the pulmonary artery (hyparterial) while the morphologically right bronchus is above the artery (eparterial). Black star = bronchus; white star = pulmonary artery.

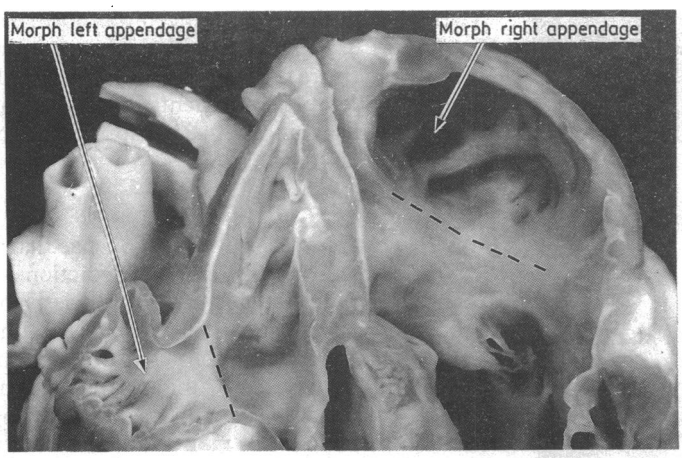


Figure 2 The normal heart is opened out to show the internal structures of the morphologically left and right atria. The tubular left atrial appendage has a narrow junction with the venous component of the atrium whereas the triangular right atrial appendage has a broad junction (dotted lines).

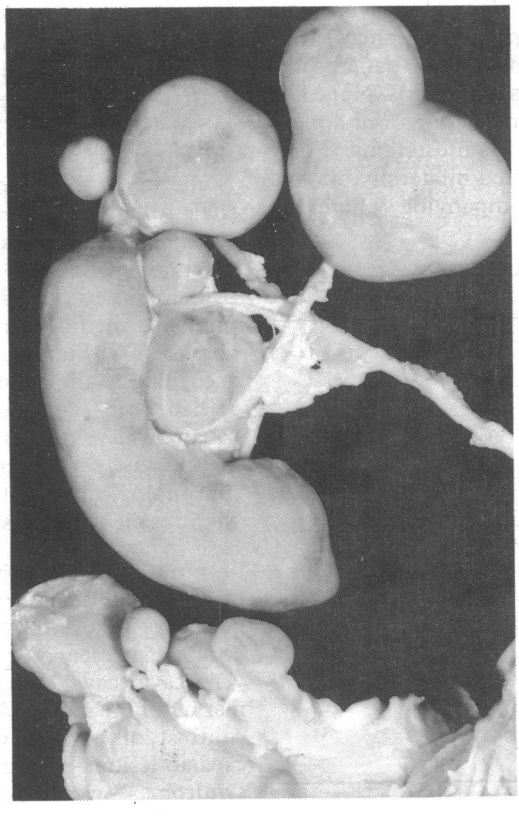
ism of the thoracic organs can have normally constructed hearts with the atria in their normal position.[1 9] But how common is this finding? And how frequently are accessory, as opposed to multiple, spleens encountered by the paediatric pathologist? These questions are vital for those who try to understand and categorise congenital malformations of the heart.[10 11] To answer them, we have reviewed the necropsy records from Children's Hospital of Pittsburgh over an 11 year period to ascertain the frequency and relation of abnormalities of the heart and thoracoabdominal organs.

Patients and methods

We examined the detailed reports on the 1158 necropsies carried out on children who died at the Children's Hospital of Pittsburgh from 1976 to 1986 inclusive. Of these reports, 116 were not included in the final analyses because the necropsy had either been exclusively thoracic or abdominal. We did not include two cases of conjoined twins because their hearts were fused.

We took information on the arrangement of the lungs, bronchi (fig 1), and atrial appendages (fig 2) from each report and noted any congenital abnormalities of the heart and great vessels. These structures were assumed to be normal if not specifically described to the contrary. The state of the spleen was recorded, as well as any evidence of abnormal arrangement of the other abdominal organs. The spleen was designated to be normal, absent, multiple (fig 3), or accompanied by accessory splenules (fig 4). Multiple spleens were recorded only when specifically described as such by the reporting pathologist. It was more common for additional splenic tissue, when noted, to be reported as accessory. The hearts of all the patients with isomerism of the atrial appendages had been retained in the Heart Museum. We re-examined these in sequential segmental fashion, making particular note of the veno-atrial connections and any associated malformations (fig 5).

Figure 3 Multiple spleens.



Results

Table 1 shows the general findings from the survey of the 1042 cases, broken down according to splenic morphology. According to the morphology of the atrial appendages, a total of eight patients (0·77%) in the overall series showed morphologically left isomerism (table 2) while 13 (1·25%) showed morphologically right isomerism (table 3). Another seven cases (0·67%) were discovered to have multiple spleens without any evidence of isomerism of the atrial appendages (table 4). There were no instances of complete absence of the spleen without left or right isomerism of the atrial appendages, but four cases had hypoplastic spleens (weighing, on average, 1·6 g). The spleen in one weighed only 0·6 g.

Five of the 116 incomplete reports are worthy of mention. Two were described as having "polysplenia", but a postmortem examination of the thoracic organs had not been performed to confirm the presence (or absence) of isomerism of the appendages. Another patient showed isomerism of the left atrial appendages and one isomerism of the right appendages, but no abdominal inspection had been carried out to record the state of the spleen. A fifth example of isomerism of the right appendages was noted in one of the conjoined twins. It is probable, therefore, that these cases would have increased the incidence of isomerism of the left and right appendages in the overall population.

Two of the 1042 cases had a mirror image atrial arrangement. Because the position of the lungs, bronchi, and abdominal organs showed appropriate mirror imagery these were all included in the category of those having normal spleens and lungs with attendant congenital heart disease.

There were 11 reports (1·06%) of accessory spleens accompanied by malrotation of the gut.

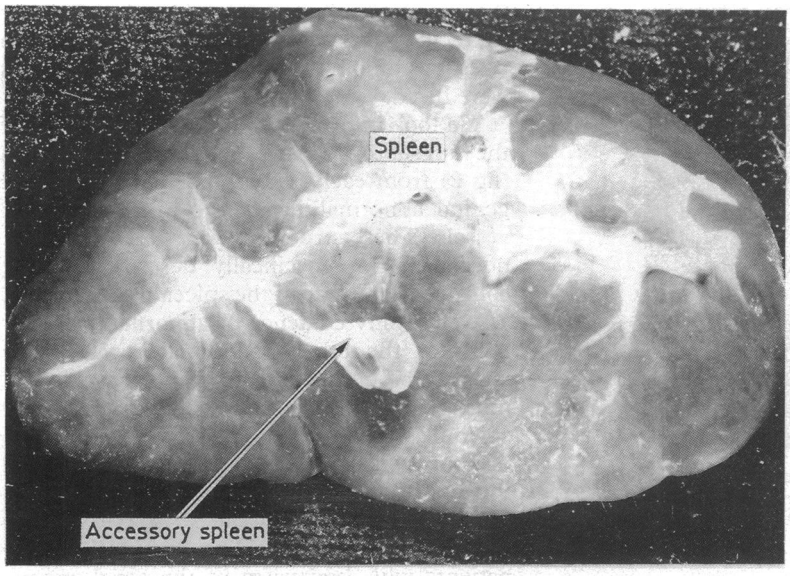


Figure 4 An accessory spleen.

One of these also had bilaterally bilobed lungs and hyparterial bronchi but all showed the usual atrial arrangement. Ten had diaphragmatic hernias, chromosomal disorders, or biliary atresia. Four cases had congenital heart disease, but not in the combinations expected for isomerism of the atrial appendages.

Isolated malrotation of the gut with a normal spleen was noted in 19 necropsies (1·82%). In 13 of these there were either diaphragmatic hernias or chromosomal disorders. Atrial arrangement was usual in all instances, with nine patients having some form of congenital heart disease. Abnormal lung lobation was mentioned twice, but neither patient had an isomeric arrangement of the bronchi.

Abnormalities of lobation of the lungs were reported in 80 cases (7·68%). Thirteen (1·25%) of these had bilaterally trilobed lungs (one case also having bilaterally eparterial bronchi). The spleen was normal in nine of these patients and the other four had accessory spleens. None had either form of isomerism of the atrial appendages or their cardiac manifestations. Forty four (4·22%) reports noted bilaterally bilobed lungs (four with accompanying bilaterally hyparterial bronchi). The spleen was normal in 35 of these patients, eight had accessory spleens, and the remaining patient had multiple spleens. None of these patients, however, had either isomerism of the left atrial appendages or the expected cardiac lesions.

Table 1 Summary of findings from 1042 necropsies

Categories		*No*	*% of total*
Cases of normal spleen:			
Normal lungs and normal heart		562	53·93
Normal lungs and abnormal heart		193	18·52
Abnormal lungs and normal heart		34	3·26
Abnormal lungs and abnormal heart*		37	3·55
	Total	*826*	*79·26*
Cases of accessory spleens:			
Normal lungs and normal heart		116	11·13
Normal lungs and abnormal heart		50	4·80
Abnormal lungs and normal heart		8	0·77
Abnormal lungs and abnormal heart		16	1·54
	Total	*190*	*18·24*
Cases of multiple spleens:			
Usual atrial arrangement		7	0·67
Left atrial isomerism		6	0·58
	Total	*13*	*1·25*
Cases of absent spleen:			
Right atrial isomerism		12	1·15
Left atrial iosomerism		1	0·10
	Total	*13*	*1·25*

*One case had right atrial isomerism.

Discussion

Our review of a large series of paediatric necropsies confirms our earlier predictions.[3 5] It is no longer reasonable to attempt to describe or diagnose congenital malformations of the heart in terms of abnormalities of the spleen. The attention given to the "splenic syndromes" has served its purpose in drawing attention to the cardiac malformations known to accompany them.

Our findings emphasise the relative constancy of the cardiac lesions themselves. Thus on the one hand there are hearts characterised by bilateral atrial appendages of right morphology. These hearts have bilateral superior caval veins, totally anomalous pulmonary venous connection, a common atrioventricular valve, and a high proportion of double inlet atrioventricular connection and an abnormal ventriculoarterial connection, usually with pulmonary stenosis or atresia.[1] On the other hand there are hearts with bilateral appendages of left morphology. These hearts characteristically have bilateral superior venae cavae (caval veins) and a common atrioventricular valve, but they have a relatively normal arrangement of their ventricular mass, coarctation rather than pulmonary atresia, and an anomalous connection of the inferior vena cava (caval vein), usually with azygos continuation.[2 8] The most accurate guide to the presence of these cardiac lesions is neither absence of the spleen nor the presence of multiple spleens. It is the existence of isomerism of the atrial appendages.[6]

When we describe "atrial isomerism", it should not be presumed that each atrium, along with its venous connections, is an isomer of the other. Far from it. The isomerism affects only the appendages, and these structures are truly isomeric. In this respect, it is the appendages that fulfil the criteria established by Van Praagh and his colleagues for determining the morphology of any cardiac chamber.[12] The principle they established[12] is called the morphologic method. In essence, it states that chambers within the heart should be identified according to their most constant components. It had been suggested that the inferior vena cava (caval vein) was the most constant atrial component, serving in this way to identify the morphologically right atrium.[13] The analysis of the present patients shows that this is not the case.

In our patients with abnormal arrangement of the abdominal organs, where it is most necessary accurately to determine atrial morphology, presence or absence of the inferior vena cava (caval vein) provides an ineffective marker of the morphologically right atrium. In contrast, all patients with the constellation of cardiac lesions previously known as "splenic syndromes" would have been correctly iden-

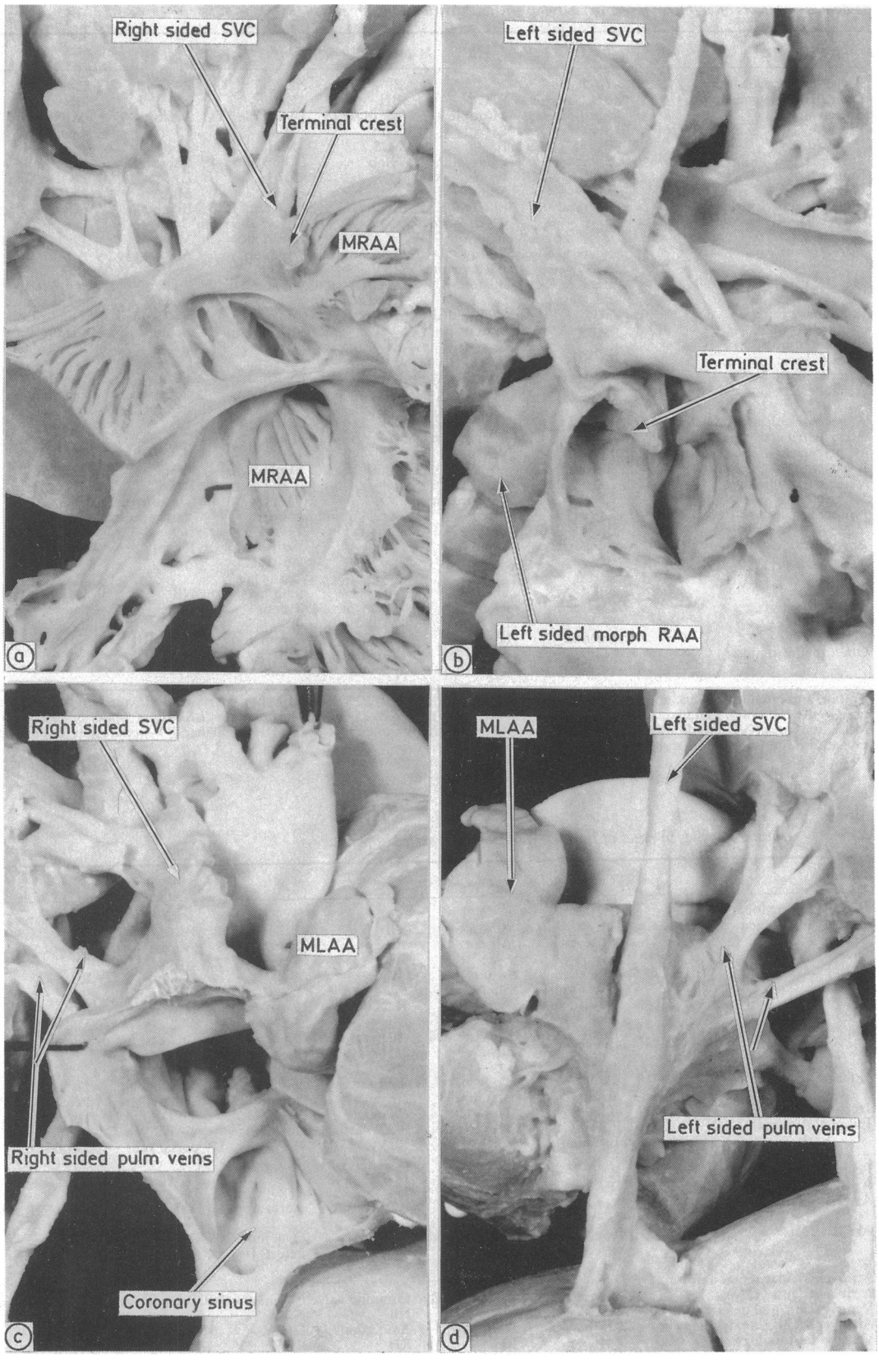


Figure 5 *The top panels show a heart with isomerism of the right atrial appendages: (a) right sided atrium, (b) left sided atrium. Note the presence of bilateral venae cavae and terminal crests. The lower panels show the internal arrangement of the atria in the presence of isomerism of the left atrial appendages: (c) right sided atrium, (d) left sided atrium. Note the presence of bilateral superior venae cavae and pulmonary veins connecting directly to the atria. MRAA, morphologically right atrial appendage; RAA, right atrial appendage; morph, morphologically; MLAA, morphological left atrial appendage; pulm, pulmonary.*

tified on the basis of isomerism of their atrial appendages. Thus the anatomy of the appendage and the arrangement of its junction with the venous atrial component provide an accurate means of distinguishing the morphologically right and left atria even in the presence of the most complex malformations of the heart. It is this criterion that best determines the arrangement of the atria. Furthermore, when the atria are abnormally formed in association with congenital cardiac anomalies, it is more accurate to describe them in terms of atrial isomerism rather than invoking considerations of malformations of the spleen.

Determining the arrangement of the atria is relatively easy for the pathologist, who holds the heart in his hands, and also for the cardiac surgeon, who can readily recognise the structure of the appendages as seen in the operating room. But what of the clinician, who is often

Table 2 Details of eight cases of isomerism of the left atrial appendages

Feature	*1*	*2*	*3*	*4*	*5*	*6*	*7*	*8*
Right/left lung lobation	2 2	2 2	2 2	2 2	2 2	2 2	1 2	2 2
Bronchial arrangement	BH	BH	BH	BH	BH	BH	BH	BH
Spleen	Multiple 5	Multiple 9	Multiple 15	Multiple 12	Multiple 4	Multiple 7	Normal	Absent
Liver	Sym	L	Sym	Sym	L	Sym	Sym	R
Gallbladder	R	L	Mid	Mid	Mid	Mid	Mid	NK
Pancreas	Short mid	Glob mid	Glob	Glob mid	Short mid	Short mid	Glob	Glob
Stomach	R	L	L	L	L	L	R	R
Gut	Mal	Mal	Mal	Mal	Mal	Mal	Mal	Mal
Superior venae cavae	Bilateral	Bilateral L to CS to R-sided atrium	Bilateral	Bilateral L to L-sided atrium atretic R	Bilateral	Bilateral atretic left	L to L-sided atrium	R to R-sided atrium
Inferior vena cava	Bilateral SVCs & azy veins	Right to R-sided atrium	Azy cont to RSVC	Azy cont to LSVC	Azy cont to LSVC	Azy cont to RSVC	Azy cont to LSVC	Azy cont to RSVC
Hepatic veins	Confl to R-sided atrium	Bilateral	2 veins to R-sided atrium	Bilateral	Bilateral	Confl to L-sided atrium	Confl to R-sided atrium	Confl to R-sided atrium
Pulmonary veins	To L-sided atrium	To R-sided atrium	Bilateral	Bilateral	To L-sided atrium	To L-sided atrium	To R-sided atrium	To L-sided atrium
Position of heart	L	L	L	L	L	L	R	L
Apex of heart	L	L	L	L	L	L	R	L
Arterial relations	Normal	Normal	Normal	Normal	Normal	Normal	Mirror image	Normal
Atrial defects	Common atrium	Common atrium	Common atrium	Common atrium	Common atrium	Common atrium	Common atrium	Common atrium
Atrioventricular septal defect	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Atrioventricular valves	Common valve	Common valve	Common valve	2 valves	Common valve	Common valve	Common valve	Common valve
Ventricular septal defect	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes
Atrioventricular connection	Ambig	Ambig	Ambig	Ambig	Ambig	Ambig	Ambig	Ambig
Ventricular topology	RH	RH	RH	LH	RH	RH	LH	RH
Ventricular arterial connection	Concordant	Concordant	DORV	DORV	Concordant	Concordant	Concordant	Concordant
Pulmonary outflow tract	Patent	Patent	Musc subpulm stenosis	Patent	Valvar pulmon stenosis	Patent	Musc stenosis	Patent
Aortic outflow tract	Subaortic stenosis	Stenosis	Patent	Subaortic obstruct by tissue tag	Patent	Stenosis	Patent	Patent
Coarctation	Nil	Shelf	Nil	Isthmal hypo	Nil	Isthmal hypo	Nil	Isthmal hypo
Arterial duct	Ligament	Patent	Ligament	Patent	Ligament	Patent	Patent	Ligament
Aortic arch	L	L	L	L	L	L	R	L
Left ventricle	Normal	Hypo	Normal	Normal	Normal	Normal	Normal	Normal
Other malformations	Nil	Nil	R juxta atrial append	Nil	Nil	Nil	RV hypo	Nil

Ambig, ambiguous; append, appendages; azy cont, azygos continuation; BH, bilaterally hyparterial; Confl, confluent; CS, coronary sinus; DORV, double outlet right ventricle; Glob, globular; Hypo, hypoplasia; L, left; LH, left-hand; Ligament, ligamentous; LSVC, left superior vena cava; Mal, malrotation; Mid, midline; Musc, muscular; NK, not known; Obstruct, obstruction; Pulmon, pulmonary; R, right; RH, right-hand; R juxta, right juxtaposition of; RSVC, right superior vena cava; Subpulm, subpulmonary; Sym, symmetrical.

Table 3 Details of thirteen cases of isomerism of the right atrial appendages

Feature	*1*	*2*	*3*	*4*	*5*	*6*	*7*	*8*
Right/left lung lobation	3 3	3 3	3 3	3 3	3 3	3 3	2 2	3 2
Bronchial arrangement	BE	BE	BE	BE	BE	BE	BE	BE
Spleen	Absent	Absent	Absent	Normal	Absent	Absent	Absent	Absent
Liver	Sym	Sym	L	Sym	Sym	R	Sym	Sym
Gallbladder	R	Mid	Mid	NK	NK	R	L	R
Pancreas	Short mid	Short mid	Glob mid	L	NK	R	R	Elong
Stomach	Mid	R	R	L	NK	R	R	NK
Gut	Mal	Mal	Mal	Normal	Mal	Mal	Mal	Mal
Superior venae cavae	Bilateral atretic right	L to L-sided atrium	Bilateral	Bilateral; L to L-sided A; atretic R	Bilateral	Bilateral	Bilateral atretic left	Bilateral
Inferior vena cava	To L-sided atrium	To R-sided atrium	To L-sided atrium	To L-sided atrium	To L-sided atrium	To R-sided atrium	To R-sided atrium	To L-sided atrium
Hepatic veins	Bilateral	To IVC	Bilateral	To IVC	To IVC	To IVC	To IVC	To IVC
Pulmonary veins	TAPVC Infradia	TAPVC to portal vein	TAPVC to RSVC	TAPVC to RSVC	TAPVC to portal vein	TAPVC R to portal L to LSVC	TAPVC to R-sided A	TAPVC to R-sided A
Position of heart	R	R	L	L	L	L	R	R
Apex of heart	R	R	L	L	L	L	R	R
Arterial relations	Aorta ant	Aorta to R	Aorta ant & right	Aorta ant	Aorta ant	Aorta to R side by side	Aorta ant	Aorta ant
Atrial defects	Oval fossa defect	Septal strand	Primum (AVSD)	Oval fossa defect	Oval fossa & primum (AVSD)	Common atrium	Oval fossa defect	Septal strand
Atrioventricular septal defect	Yes	Nil	Yes	Nil	Yes	Nil	Nil	Nil
Atrioventricular valves	Common valve	Common valve	Common valve	Common valve	Common valve	Common valve	Common valve	Common valve
Ventricular septal defect	Yes (AVSD)	Yes	Nil	Nil	Yes	Nil	Yes	Nil
Atrioventricular connection	Ambiguous	DIRV	Ambiguous	DIRV	Ambiguous	DILV	DILV	DIIV
Ventricular topology	LH	RH	RH	RH	RH	LH	LH	NA
Ventricular arterial connection	Discordant	DORV	DORV	Single outlet Ao from RV	Single outlet Ao from RV	Concordant	Single outlet Ao from RV	Single outlet Ao from IV
Pulmonary outflow tract	PA	Subpulm stenosis	Subpulm stenosis	PA	PA	PA	PA	PA
Aortic outflow tract	Patent	Patent	Patent	Patent	Patent	Patent	Patent	Patent
Coarctation	Nil	Nil	Nil	Nil	Nil	Nil	Nil	Nil
Ductus arteriosus	Patent to confl PAs	Patent	Ligament	R-sided patent	R-sided patent	R-sided patent	R-sided patent	Ligament
Aortic arch	R	R	R	L	L	L	R	R
Left ventricle	Hypo	Slit-like	Hypo	Slit-like	Normal	Dominant	Normal	
Other malformations	Nil	Nil	Imperf LAVV	Nil	Nil	RV grossly hypo	Rudimentary RV	Solitary IV

Ant, anterior; Ao, aorta; AVSD, atrial or ventricular component of atrioventricular septal defect; BE, bilaterally hyparterial; DIIV, double inlet indeterminate ventricle; DILV, double inlet left ventricle; DIRV, double inlet right ventricle; Elong, elongated; IVC, inferior vena cava; Imperf LAVV, imperforate left atrioventricular valve; Infradia, infradiaphragmatic; IV, solitary and indeterminate ventricle; L-sided A, left-sided atrium; NA, not applicable; PA, pulmonary atresia; PAs, pulmonary arteries; R-sided A, right-sided atrium; RV, right ventricle; TAPVC, totally anomalous pulmonary venous connection.

Table 4 Details of seven cases of multiple spleens and usual atrial arrangement

Case No	*Right lung lobation*	*Left lung lobation*	*Bronchial arrangement*	*No of spleens*	*Abnormal abdominal organs*	*Heart defects*	*Azygos continuation of inferior vena cava*	*Other malformations*
1	3	2	Usual	6	Globular pancreas, absent gallbladder	Nil	No	Biliary atresia
2	3	2	Usual	7	Globular midline pancreas, malrotated gut	ASD, Ao-pulmonary window	No	Biliary atresia
3	3	2	Usual	6	Malrotated gut	Nil	No	Pneumococcal septicaemia
4	3 + Acc	2 + Acc	Usual	4	Malformed stomach and pancreas	POF, ductus arteriosus, LSVC to CS	Yes	Diaphragmatic hernia
5	2	2	Usual	17	Absent gallbladder, short pancreas, malrotated gut	Nil	No	Biliary atresia
6	2	2	Bilat hypart	5	Globular midline pancreas, partially malrotated gut	Nil	Yes	Biliary atresia
7	2	1	Usual	9	Malformed pancreas, malrotated gut	POF	No	Arnold-Chiari malformation

Acc, accessory lobe; ASD, atrial septal defect; Ao-pulmonary, Aorto-pulmonary; Bilat hypart, bilaterally hyparterial; LSVC to CS, left superior vena cava to coronary sinus; POF, patent oval foramen.

unable accurately to distinguish the morphology of the appendages and the nature of their junctions with the venous components. It is recognised that inferences must be made in these circumstances to permit the paediatric cardiologist to assess atrial arrangement, although it has been suggested that cross sectional echocardiography may, in skilled hands, permit direct recognition of the appendages. Similarly, if injections of contrast material are made into the atrial appendages, cineangiography permits direct diagnosis of atrial isomerism. None the less, if inferences are to be made, our analysis shows that bronchial morphology[14] is the best guide to the existence of isomerism of the atrial appendages. All but one case with atrial isomerism also had bronchial isomerism. The exception was a case of right atrial isomerism with usual bronchial arrangement. Five patients, however, were reported to have isomeric bronchi with usual atrial arrangement in the absence of any severe cardiac lesions. One of these cases was recently highlighted by Devine *et al*,[15] but our present analysis shows them to be more frequent than we thought. Despite these "false positives", bronchial anatomy remains by far the best inferential guide to the existence of the cardiac syndromes previously linked with (and named according to) splenic malformations. We found as many patients with multiple spleens with normal hearts as with isomerism of the left atrial appendages (previously described as "polysplenia"). Those with multiple spleens and usual atrial arrangement were usually examples of a syndrome known to exist with biliary atresia.[9] Their presence in such numbers in our series may reflect Pittsburgh's role as a major centre for liver transplantation. That, however, does not diminish the finding that multiple spleens can no longer be regarded as an accurate pointer to the presence of a specific syndrome of congenital heart disease. Although no patients without cardiac lesions had no spleen, four had such gross hypoplasia of splenic tissue that it was questionable whether the organ would have been demonstrable by clinical diagnostic techniques. Furthermore, one patient with isomeric right appendages had a normal spleen while another patient with isomeric left appendages had no spleen.

The lesions previously correlated with splenic malformations are best diagnosed in terms of isomerism of the atrial appendages.[6] If diagnosis has to be made by inference, this will be achieved most accurately by referring to the pattern of bronchial morphology.[14] Splenic malformations are insufficiently well correlated with anomalies of the heart to justify their use either as an inferential guide or as a platform for nomenclature.

9	*10*	*11*	*12*	*13*
3 3	2 3	3 2	1 1	3 2
BE	Mirror image	BE	BE	BE
Absent	Absent	Absent	Absent	Absent
Sym	Sym	L	Sym	Sym
L	L	L	NK	L
NK	R	R	NK	R
L	R	R	NK	L
Mal	Mal	Mal	Normal	Mal
Bilateral	Right to R-sided atrium	Bilateral	Bilateral	Left to L-sided atrium
To L-sided atrium	L to L-sided atrium	To L-sided atrium	To R-sided atrium	To L-sided atrium
Bilateral	To IVC	Bilateral	To IVC	To IVC
TAPVC to portal vein	TAPVC to portal vein	TAPVC to R-sided A	TAPVC to R-sided A	TAPVC to R-sided A
R	R	R	R	R
R	R	R	R	R
Aorta ant & left	Aorta ant & left	Aorta ant & left	Aorta ant & left	Aorta ant
Primum (DIRV)	Septal strand	Primum (DILV)	Septal strand	Common atrium
Nil	Yes	Nil	Nil	Nil
Common valve	Common valve	Common valve	Common valve	Common valve
Nil	Yes	Yes	Yes	Yes
DIRV	Ambiguous	DILV	DILV	DILV
LH	LH	LH	LH	LH
DORV	DORV	Discordant	Single outlet Ao from RV	Single outlet Ao from RV
Subpulm stenosis	Subpulm stenosis	Severe subpulm stenosis	PA	PA
Patent	Patent	Patent	Patent	Patent
Nil	Nil	Nil	Nil	Nil
R-sided patent	R-sided patent	Patent	Patent	R-sided patent
R	R	R	L	R
Slit-like	Normal	Normal	Normal	Normal
Nil	Nil	RV hypo	RV hypo	RV hypo

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