Double inlet left ventricle *Straddling tricuspid valve*¹

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Eleven cases of double inlet left ventricle have been presented. Double inlet left ventricle is characterized by the tricuspid valve opening partly (partial type) or completely (complete type) into the morphological left ventricle. The morphological right ventricle is usually hypoplastic but may be normally formed. Even when the right ventricle is hypoplastic, both the sinus and the outflow portions of the right ventricle are recognizable.

Double inlet left ventricle has been classified into two groups according to whether the ventricles are normally placed (noninverted; concordant with the atria) or inverted (discordant with the atria.) This classification indicates the anatomical relation of the atrioventricular valves and the ventricles. When the ventricles are non-inverted, a right-to-left shunt results from the communication of the right atrium with the left ventricle. On the other hand, with inverted ventricles a left-to-right shunt results from the left atrium communicating with the inverted left ventricle. Further details in circulation depend upon the great vessel-ventricular relations.

Clinically, the patients presented with cyanosis when pulmonary stenosis was present and with congestive cardiac failure when pulmonary stenosis was absent.

The electrocardiogram is helpful in suggesting the presence of inverted or noninverted ventricles. Right and left atriograms are essential for the diagnosis of straddling tricuspid valve in noninverted and in inverted ventricles, respectively.

Straddling tricuspid valve is an anomaly wherein the tricuspid valve overrides the ventricular septum and communicates with both ventricles. This condition may be considered as one form of double inlet left ventricle. In another form of the latter condition the tricuspid valve opens entirely into the left ventricle, as does the mitral valve. The subject of double inlet left ventricle and straddling tricuspid valve have received major attention only in recent years. The subject is somewhat complex in that variations in the positions of the ventricles and of the great arteries may be associated and mitral atresia may be present.

Because of the foregoing and the fact that a limited number of cases have been reported, it appears

appropriate to add to the published reports a description of additional cases so as to provide additional material for a more comprehensive understanding of double inlet left ventricle.

Eleven cases with double inlet left ventricle formed the basis for this study. Necropsy had been done in all, and 10 specimens were available for re-evaluation. Clinical and radiological findings were available in all, and the electrocardiograms in 10 cases.

Definition of terms

Inversion

The term inversion indicates a mirror image of the usual with regard to right and left orientation. Anteroposterior relations are not altered. Inversion may involve the entire body, as in total situs inversus, or it may affect only portions of the cardiovascular system. Inversion may be confined to the ventricles, including atrioventricular valves ('isol-

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ated ventricular inversion') or great vessels, while the atria and other organs are normally positioned.

Thus, ventricular inversion implies that the morphological right ventricle is left-sided and the morphological left ventricle, right-sided. The term non-inverted applies to normal or usual relations. The atrioventricular valves may be considered part of the inflow portion of the ventricles so that the natural valve for the left ventricle is the mitral and for the right ventricle the tricuspid. If the ventricles are inverted, so are the atrioventricular valves. This means that with isolated ventricular inversion the anatomical tricuspid valve lies between the left atrium and the anatomical right ventricle, while the mitral valve lies between the right atrium and the right-sided anatomical left ventricle.

Transposition of great vessels

Transposition of the great vessels is defined as an abnormal external relation of the great vessels in that the aorta occupies a position more anterior than normal and the great vessels lie parallel to each other. The aorta arises from the morphological right ventricle and the pulmonary trunk from the morphological left ventricle. In addition to transposition, the great vessels may show inversion or non-inversion. If the transposed aortic valve lies to the right of the pulmonary valve, there is noninversion or d-transposition. If the transposed aortic valve lies to the left of the pulmonary valve and the ascending aorta sweeps along the left side of the pulmonary trunk, there is inverted or l-transposition.

Double inlet left ventricle

Double inlet left ventricle may be designated as either of the partial type or of the complete type.

The partial form of double inlet left ventricle involves either non-inverted or inverted ventricles and is characterized by the tricuspid valve overriding (straddling) a ventricular septal defect and opening partly into the morphological left ventricle and partly into the morphological right ventricle (Fig. 1 and 2). The mitral valve may either be atretic or present. If present, the mitral valve opens entirely into the morphological left ventricle. The morphological right ventricle may be hypoplastic or well formed. However, the inflow (sinus) and the outflow portions of the right ventricle are both recognizable. Excluded from the definition of double inlet left ventricle are cases of common atrioventricular valve.

The complete form of double inlet left ventricle is that wherein the tricuspid valve opens entirely

FIG. 1 Case 3. Interior of the noninverted left ventricle (L.V.). The mitral valve (M.) and part of the tricuspid valve (between arrows) communicate with this chamber. The aorta (A.) arises from the left ventricle.

into the morphological left ventricle which may be inverted or non-inverted. This is to be distinguished from the more common state of common ventricle. In the latter, the sinus portion of the right ventricle is not formed, while in those cases called double inlet left ventricle there is an identifiable right ventricular sinus.

Anatomical types

Each of the 11 cases of double inlet left ventricle to be described showed certain features in common. These were situs solitus and normal pulmonary venous and systemic venous connexions, as well as absence of any splenic anomaly.

In 10 of the 11 cases the partial type of double inlet left ventricle was present and in one, the complete type. In the following anatomical classification, primary subdivision will be on the basis of the pre-

FIG. 2 Case 6. a) Interior of the right atrium (R.A.) and inverted morphological left ventricle (L.V.). The fossa ovalis (F.O.) identifies the right atrium (R.A.). The inverted mitral valve (M.) lies between the right atrium and the morphological left ventricle. The inverted tricuspid valve (T.) straddles the ventricular septum and communicates, in part, with the morphological left ventricle. b) The left atrium (L.A.) and the inflow portion of the inverted morphological right ventricle (R.V.). The tricuspid valve also communicates in part with the inverted right ventricle.

sence or absence of ventricular inversion and of transposition of the great vessels.

Double inlet noninverted left ventricle (group I)

There were 5 specimens wherein the ventricles were non-inverted and were concordant with the atria. In each, the double inlet left ventricle was of the partial type. In 3 of these, the great vessels were normally related and in 2 the great vessels exhibited d-transposition.

The mitral valve was atretic in one (Case 4) and present in each of the remaining 4 cases. When present, the mitral valve opened entirely into the left ventricle. Through a ventricular septal defect, the tricuspid valve straddled the ventricular septum, opening partly into the right and partly into the left ventricle (Fig. 3 and 4). The septal leaflet alone or

the septal and either the posterior or the anterior leaflets of the tricuspid valve were the straddling ones. Chordae from these leaflets attached to the left ventricular side of the ventricular septum or to papillary muscles of the left ventricle. The right ventricle was hypoplastic in each case (Fig. 3a) except the one (Case 2) in which aortic atresia was also present (Fig. 3b). In this case the right ventricle was larger than the left ventricle.

The 5 cases with non-inverted (concordant) ventricles could be subdivided into two groups according to the position and origin of the great vessels. In 3 cases (Cases 1 to 3) the great vessels were normally interrelated and arose normally (Fig. 3). (In Case 2, aortic atresia was present.) In the other 2 cases, the great vessels exhibited d-transposition and the aorta arose from the right ventricle, while the pulmonary trunk arose from the left ventricle (Fig. 4). The one example of mitral

FIG. 3 Diagrammatic portrayal of double inlet, noninverted left ventricle with normally related great vessels. *a*) Case 1. The right ventricle (R.V.) is hypoplastic and the left ventricle (L.V.) large. The tricuspid valve (T.) straddles the ventricular septum. A, aorta; P.T., pulmonary trunk. *b*) Case 2. The right ventricle (R.V.) is larger than the left ventricle (L.V.). The aortic valve is atretic and the ascending aorta hypoplastic. The systemic flow is derived through the persistent ductus arteriosus (P.D.A.). The mitral valve shows a parachute deformity, its leaflets being attached to one papillary muscle. The tricuspid valve (T.) straddles the ventricular septum.

atresia associated with non-inverted ventricles was one of the two with d-transposition (Case 4) (Fig. 4a and 5).

Double inlet inverted left ventricle (group 2)

Six specimens were characterized by the presence of ventricular inversion (ventricles discordant with the atria). In 4 of these, l-transposition was associated and in each of these the double inlet was of the partial type. In the fifth case (Case 10), also with the partial type of double inlet left ventricle, the great vessels occupied a position of d-transposition, though the aorta arose from the inverted right ventricle and the pulmonary trunk from the inverted left ventricle. The transposition in this is designated as atypical.

The sixth case with ventricular inversion (Case 11) had unusual features including normally related great vessels and will be described separately after a consideration of the first five cases of this group. This case was the only example of the complete type of double inlet left ventricle.

Transposition of the great vessels (l-type in 4 and atypical (Case 10) in 1) was present in 5 of the 6 cases with ventricular inversion. In these, the right atrio-ventricular valve was considered to be the morphological mitral valve and the left, the tricuspid. In one of these cases, the right-sided mitral valve was atretic (Case 10), while in the remaining 4 the mitral valve was present and opened entirely into the right-sided morphological left ventricle (Fig. 2a, and 6a and b). In each of these 5, the anterior and the septal leaflets of the left-sided morphological tricuspid valve straddled the ventricular septum so that this valve opened partly into each ventricle. In 4 specimens, chordae from the 'straddling' leaflets attached to a papillary muscle of the morphological left ventricle and in one specimen to the septal wall of the morphological left ventricle. The morphological right ventricle was hypoplastic in each of the 5 cases (Fig. 2b).

In 4 of the first 5 cases in this group the great vessels were transposed and in 4 this took the form of l-transposition, yielding an anatomical state

FIG. 4 Double inlet noninverted left ventricle with *d*-transposition of great vessels. *a*) Case 4. The tricuspid valve (T.) straddles the ventricular septum and the mitral valve is atretic. *b*) Case 5. The aortic arch is right-sided with mirror image branching. There is also left-sided juxtaposition of atrial appendages.

FIG. 5 Case 4. *a*) External view of the heart and great vessels showing *d*-transposition. *b*) Interior of the right atrium (R.A.), left ventricle (L.V.), and straddling tricuspid valve (T.). The right ventricle (R.V.), which is out of view, was hypoplastic. *c*) Interior of the left atrium (L.A.) showing site of atretic mitral valve (arrow). The probe is in interatrial ostium II.

FIG. 6 a) Case 5. Double inlet inverted left ventricle (L.V.) with l-transposition of great vessels. The mitral valve (M.) enters the inverted morphological left ventricle (L.V.) and the morphological tricuspid valve (T.) straddles the ventricular septum. b) Case 7. Pulmonary valvular atresia is present. A persistent ductus arteriosus (P.D.A.) provides the pulmonary blood flow. c) Case 10. Atypical transposition. Though the great vessels show a status of d-transposition, their connexions are atypical. The aorta arises from the inverted right ventricle and the pulmonary trunk from the inverted left ventricle. The mitral valve is atretic. Tubular hypoplasia of the aortic arch is present.

classical for corrected transposition (Fig. 6a and b). In the fifth case (Case 10), the great vessels showed atypical transposition. Though the aorta arose from the left-sided morphological right ventricle anterior to the pulmonary trunk, it then ascended to the right of the pulmonary trunk (Fig. 6c). The latter vessel arose from the right-sided morphological left ventricle. This case is viewed as an example of atypical transposition of the great vessels anatomically, but functionally like that of corrected transposition.

Ventricular inversion in the absence of transposition characterized the eleventh case of this series. In this case, both atrioventricular valves opened entirely into the inverted (right-sided) morphological left ventricle resulting in the complete type of double inlet left ventricle. The left atrioventricular valve was small. The anterior leaflet of the right atrioventricular valve was attached to the superior rim of a large muscular ventricular septal defect. An anomalous muscle bundle was present in the inverted morphological left ventricle. The inverted morphological right ventricle was well

formed and communicated with the morphological left ventricle through one large and several small muscular ventricular septal defects. The aorta arose from the morphological inverted left ventricle and the pulmonary trunk from the morphological inverted right ventricle (Fig. 7).

Associated anomalies

The associated anomalies observed in our cases are summarized in Table 1. Obstruction to pulmonary blood flow was present in 3 cases. In 2 of these, it took the form of subpulmonary stenosis and in one pulmonary valvular atresia. Obstruction to outflow from the left atrium occurred in 2 cases and was represented by parachute mitral valve (one case) and mitral atresia (one case). In an additional case, mitral atresia was present in association with inverted ventricles and caused obstruction to outflow from the right atrium.

Aortic valvular atresia associated with coarctation of the aorta was present in one case; in 4 other cases, obstruction in the aorta was present in the

FIG. 7 Case 11. Double inlet inverted left ventricle of the complete type. Both the right atrioventricular valve (R.A.V.) and the stenotic left atrioventricular valve (L.A.V.) enter the inverted left ventricle (L.V.). The anterior leaflet of the right atrioventricular valve is attached to the superior rim of the large muscular ventricular septal defect. (Smaller ventricular septal defects not shown.) The aorta (A.) arises from the inverted left ventricle and the pulmonary trunk (P.T.) from the inverted right ventricle. The external relation of the great vessels was normal.

form of classical coarctation in one case and tubular hypoplasia of the aortic arch in 3.

Clinical findings

The ages of the 11 patients (6 boys and 5 girls) in this series varied from 5 days to 10 years. Only 3 patients were older than 5 months, their ages being 2½, 3½, and 10 years, respectively. The family history was significant in only one patient (Case 3) whose cousin had been found at necropsy to have aortic valvular stenosis with left ventricular endocardial fibroelastosis. Each of the patients who survived beyond two weeks showed poor growth. Because of similar clinical features, findings of groups 1 and 2 will be presented together.

One baby was blue at birth and 3 others developed cyanosis within the first week of life. One infant who was acyanotic at 6 weeks of age developed mild cyanosis by the age of 5½ months and moderately severe cyanosis by 9 months (Case 3). Pathological study showed that obstruction to pulmonary blood flow was present in 3 of the 5 cases with cyanosis. In

2 of these, there was pulmonary stenosis (Cases 3 and 5) and, in one case, pulmonary atresia (Case 7).

Congestive cardiac failure was present in 6 of the 8 cases without obstruction to pulmonary blood flow. Two of these were investigated in infancy because of failure to respond to anticongestive measures. A persistent ductus was divided in both. One of these (Case 1) survived to the age of 3½ years. In the other (Case 6), resection of coarctation of the aorta and banding of the pulmonary trunk were also performed and the patient survived to the age of 10 years.

Hypoxic spells were present in only one patient (Case 3). This patient initially presented with features of a ventricular septal defect with left-to-right shunt. Over the course of several months, signs of increasing severity of pulmonary stenosis developed.

The presenting symptoms in one patient (Case 11), the solitary case with Down's syndrome in this series, were related to an associated tracheo-oesophageal fistula.

Physical examination disclosed extracardiac anomalies in 3 patients. In addition to the one case with Down's syndrome, one showed features of trisomy-18, with multiple musculoskeletal anomalies including rocker-bottom feet, pes cavus, abnormal facies, ears fused to scalp, absent right clavicle, and harelip (Case 2). In the third patient (Case 3) a lumbosacral myelomeningocele was successfully repaired in infancy, the patient surviving to the age of 2½ years.

Physical examination of the cardiovascular system showed nonspecific findings. Cardiomegaly and congestive failure were present in 6 patients.

The first cardiac sound was unremarkable in each of the 11 patients. The second sound was variously described as normal, single, or split with accentuated pulmonary component. Except for one patient (Case 2), a systolic murmur was heard in each. It varied in intensity from grade 2 to 4/6 and was variably described as pansystolic or ejection systolic. A delayed diastolic rumbling murmur at the apex was described in 2 patients. In the one patient with pulmonary atresia (Case 7), a murmur was not heard initially but by the age of 11 days a continuous machinery-type murmur was audible.

Electrocardiographic findings

Electrocardiograms were available for re-evaluation in 10 cases. The frontal plane QRS axes are shown in Fig. 8. Electrocardiograms were available in 4 of the 5 cases with non-inverted ventricles. In these, the frontal plane QRS axis was to the left in 3 and normal (+30°) in the fourth case. Evidence for

TABLE I Summary of clinical and pathological findings in the 11 cases of double inlet left ventricle studied

Case No.	Age	Sex	Pulmonary stenosis	Great vessel relation	Cyanosis	Congestive cardiac failure	Cardiac enlargement	Pulmonary vasculature	Axis QRS	V4R or V1	V6	Associated anomalies
<i>Noninverted ventricles</i>												
1	3½ yr	F	o	Normal	o	+	+	Increased	-60	rsr's'	qRs	Persistent ductus arteriosus; supraventricular ring left atrium
2	21 dy	F	o	Normal	+	o	+	Increased	—	—	—	Aortic atresia; parachute mitral valve; coarctation aorta; persistent ductus arteriosus; trisomy-18 with multiple congenital anomalies
3	2½ yr	M	+	Normal	+	o	o	Diminished	+30	rS	R	Bicuspid pulmonary valve; myelomeningocele
4	5 mth	M	o	d-transposition	o	+	+	Increased	-105	qR	RS	Mitral atresia
5	1 mth	F	+	d-transposition	+	o	o	Diminished	-60	rS	qRs	Right aortic arch with mirror image branching; juxtaposition of atrial appendages
<i>Inverted ventricles</i>												
6	10 yr	F	o	l-transposition	o	+	+	Increased	+120	qR	Rs	Coarctation of aorta
7	3½ mth	M	+*	l-transposition	+	o	o	Diminished	+110	R	RS	Persistent ductus arteriosus
8	6 wk	M	o	l-transposition	o	+	+	Increased	+105	qR	RS	Persistent ductus arteriosus; tubular hypoplasia aortic arch; absent coronary sinus; atrial septal defect
9	2 mth	F	o	l-transposition	o	+	+	Increased	+80	qR	RS	Tubular hypoplasia of aortic arch; persistent ductus arteriosus
10	6 wk	M	o	Atypical transposition	o	+	+	Increased	+110	R	RS	Tubular hypoplasia of aortic arch; mitral atresia
11	5 dy	M	o	Normal	+	o	+	Increased	-75	qR	rS	Mitral stenosis; ventricular septal defect; atrial septal defect; persistent ductus arteriosus; Down's syndrome; tracheoesophageal fistula

* Pulmonary valvular atresia

I Non-inverted ventricles

- A) Normally related great vessels
- ▲ B) Complete (d) transposition

II Inverted ventricles

- A) Corrected (l) transposition
- B) Normally related great vessels

FIG. 8 The mean frontal plane QRS axis in the electrocardiogram from 10 patients with double inlet left ventricle. Of the 5 cases with inverted ventricles and l-transposition, 4 showed right axis deviation and the remaining case an axis of $+80^\circ$.

atrial hypertrophy was present in 2 electrocardiograms, one showing right and the other biatrial hypertrophy. Isolated left ventricular hypertrophy was present in the electrocardiograms of 2 patients, each with obstruction to pulmonary blood flow. Of the 2 patients without obstruction to pulmonary blood flow, the electrocardiogram showed right ventricular hypertrophy in one and biventricular hypertrophy in the other.

Among the 5 cases with inverted ventricles and transposed great vessels, the frontal plane QRS axis was to the right in 4 and normal ($+80^\circ$) in 1 case. In contrast, in the solitary case of ventricular inversion without transposition (Case 11) the QRS axis was to the left (-75°). Isolated right atrial hypertrophy was present in 4 of the 6 electrocardiograms. None showed left atrial hypertrophy. Biventricular hypertrophy was present in 4 and isolated right ventricular hypertrophy in 2 electrocardiograms. Both patients with obstruction to pulmonary blood flow (pulmonary atresia and sur-

gical banding of the pulmonary trunk) showed biventricular hypertrophy. A qR morphology of the QRS complex was present over the right praecordial leads in 4 electrocardiograms, the other 2 showing 'pure R' complexes. A 'Q' wave was not present over the left praecordial leads in any of the electrocardiograms from this group.

Radiological findings

No diagnostic features were obtained from standard radiological studies. This applied as well to the group with inverted ventricles, none of whom showed the classical radiological features of l-transposition.

The cardiac size varied from normal to conspicuous enlargement with nonspecific configuration in members of both major groups. In general, the cardiac size was enlarged with increased vascularity in those without pulmonary stenosis and normal with diminished pulmonary vascularity in those with pulmonary stenosis.

Cardiac catheterization and angiocardiology

Cardiac catheterization was performed in 5 of the 11 patients and angiocardigraphic studies in 6 patients but these investigations did not lead to a correct diagnosis of double inlet left ventricle in any of the cases. Among the 6 patients in whom angiocardigrams were done, atriograms were performed in only 2 patients, each with non-inverted ventricles. In one of the latter patients (Case 3) a right atriogram done after attempted surgical closure of the ventricular septal defect and resection of the right ventricular outflow obstruction disclosed two streams of contrast material leaving the right atrium. One of the streams opacified the hypoplastic right ventricle and the other the left ventricle. These diagnostic features were interpreted as those of a residual ventricular septal defect.

In another patient (Case 5), the right atriogram opacified only the left ventricle, which was large, whereas a left atriogram opacified both ventricles. These observations cannot be explained by the anatomical findings.

In one patient (Case 6) with inverted ventricles, the laevophase of a pulmonary arteriogram showed interesting features. After opacification of the left atrium, there was opacification of both ventricles. The hypoplastic inverted left-sided right ventricle was mistaken for the left atrial appendage while the image of the inverted right-sided left ventricle was interpreted as that of a single ventricle (Fig. 9).

FIG. 9 Case 6. Laevophase of pulmonary angiogram. The hypoplastic inverted right ventricle (R.V.) was interpreted as the left atrial appendage and the inverted left ventricle (L.V.) considered to be a single (common) ventricle. Restudy of this film along with the pathological specimen (Fig. 2 and 6a) indicates that the features shown are characteristic for double inlet left ventricle in the presence of inverted ventricles. L.A. = left atrium.

Comment

Double inlet left ventricle is a rare anomaly. Anatomically, it is characterized by opening of the tricuspid valve partially or completely into the left ventricle.

The primitive atrioventricular canal, from which both atrioventricular valves develop, faces that part of the primitive ventricle from which the left ventricle is to form. Normally, as the right ventricle is derived as an out-pouching of the primitive ventricle, the atrioventricular canal shifts towards the right so that the tricuspid valve faces the right ventricle. Arrests at various stages in development of this process may be observed in specimens as follows. The right ventricle may be well developed or hypoplastic while the tricuspid valve straddles the ventricular septum. A more deficient state of development results in a hypoplastic right ventricle but with a developed sinus while the tricuspid valve opens into the left ventricle. The most primitive state is that in which only the infundibulum of the right ventricle has developed and the tricuspid valve opens into a common ventricle. We have designated the states illustrated in Fig. 10a and b as the partial type of double inlet left ventricle and that in

Fig. 10c as the complete type. When the right ventricular sinus is not formed (Fig. 10d), classical single ventricle may be identified.

Our definition of double inlet left ventricle conforms with the views of Mehrizi *et al.* (1966), as well as of Liberthson *et al.* (1971). However, while de la Cruz and Miller (1968) and Van Mierop (1973) use the term double inlet left ventricle to include cases like ours, they have excluded cases associated with mitral atresia. On the other hand, they have included single (common) ventricle of the left ventricular type as examples of double inlet left ventricle. Van Praagh, Ongley, and Swan (1964) have used the term single ventricle type A both for double inlet left ventricle as we have defined it as well as for common ventricle of the left ventricular type.

We have classified our cases of double inlet left ventricle into two types: those with normally positioned ventricles (non-inverted) (group 1) and those with inverted ventricles (group 2). This classification is used for two reasons. First, it indicates the anatomical position of the ventricles and the connexions of the atrioventricular valves. Secondly, it also indicates the haemodynamic effect resulting from straddling tricuspid valve. With

FIG. 10 a) Partial type of double inlet left ventricle with a large right ventricle. b) Partial type of double inlet left ventricle with a small right ventricle. c) Complete type of double inlet left ventricle with a well-developed right ventricular sinus. d) Common (single) ventricle of the left ventricular type. The right ventricle is represented by the infundibulum.

noninverted ventricles, a straddling tricuspid valve would result in some of the systemic venous blood stream entering the left ventricle as an obligatory right-to-left shunt into the left ventricle. In contrast, with inverted ventricles, the left-sided tricuspid valve straddles the ventricular septum and results in an obligatory left-to-right shunt into the right-sided morphological left ventricle. In the absence of obstruction to pulmonary or systemic blood flow or abnormally attached great vessels, the main determinant of the haemodynamic abnormality would, therefore, be the presence or absence of ventricular inversion.

Two types of great vessel relations were observed in each of the two types of ventricular relations. With non-inverted ventricles, the great vessels were either normally related or exhibited the relation of d-transposition. In the group with inverted

ventricles, the great vessels usually showed an external relation as in l-transposition.

In one case, however, the aorta arising from the inverted right ventricle ascended to the right of the pulmonary trunk as is seen in d-transposition. In none of our cases of double inlet left ventricle was double outlet right ventricle coexistent. Mehrizi *et al.* (1966), however, reported features of one such case occurring in an example without ventricular inversion (double inlet left ventricle with double outlet right ventricle) (Fig. 11a). Such a state is theoretically possible in association with inverted ventricles as well (Fig. 11b). For this reason, we have included this subgroup of great vessel relation in our classification (Table 2).

Normally related great vessels with ventricular inversion are rare. Of the 50 specimens of heart with ventricular inversion available to us, our Case 11

FIG. II a) Double inlet noninverted left ventricle (L.V.) with double outlet right ventricle (R.V.). The tricuspid valve (T.) straddles the ventricular septum. Aorta (A.) and pulmonary trunk (P.T.) arise from the right ventricle. There is discontinuity between the semilunar and atrio-ventricular valves. b) Double inlet inverted left ventricle with double outlet right ventricle. Both great vessels arise from the inverted morphological right ventricle which is on the left side. There is semilunar-atrioventricular valve discontinuity.

TABLE 2 Classification of double inlet left ventricle

1) Non-inverted ventricles
a) Normally related great vessels
b) d-transposition
c) Origin of both great vessels from right ventricle
2) Inverted ventricles
a) l-transposition
b) Normally related great vessels
c) Origin of both great vessels from right ventricle

is the only case in which the pulmonary trunk arose from an inverted right ventricle and the aorta from the inverted left ventricle. Rare examples of such a combination have been reported (Van Praagh and Van Praagh, 1967).

In our material the clinical profile, electrocardiograms, and radiological findings did not provide features specific for a diagnosis of double inlet left ventricle. The clinical features depend on the presence or absence of pulmonary stenosis. In spite of an obligatory right-to-left shunt in the group with non-inverted ventricles, only 2 of the 5 patients had cyanosis initially and one developed it subsequently,

seemingly on the basis of a relative increase in pulmonary stenosis. Obstruction to pulmonary blood flow was thus present in 2 of the 3 patients with cyanosis. In the group with inverted ventricles and an obligatory left-to-right shunt, only the one patient with pulmonary atresia was cyanotic. Only minimal cyanosis was noted in another patient without obstruction to pulmonary blood flow. The presence or absence of cyanosis in patients with double inlet left ventricle is thus related to pulmonary stenosis. In the absence of pulmonary stenosis, the patients presented predominantly with congestive cardiac failure for which the associated ventricular septal defect was mainly responsible.

Certain electrocardiographic findings need to be emphasized. Left axis deviation of the frontal plane QRS axis was present in 3 of the 4 patients with non-inverted ventricles and was associated with right, left, or biventricular hypertrophy. Thus, double inlet left ventricle should be included in the differential diagnosis of cyanotic or acyanotic patients with congenital cardiac disease who exhibit left axis deviation. In 4 of the 6 patients with inverted ventricles electrocardiograms showed right axis deviation, and in one, an axis of $+80^\circ$, with

right or combined ventricular hypertrophy. Of the 6 electrocardiograms, 4 were typical of those seen in l-transposition of great vessels with ventricular inversion, in having 'Q' waves over the right praecordial leads, while the 'Q' waves were absent over the left praecordial leads. Thus, double inlet left ventricle should be considered in the differential diagnosis of patients whose electrocardiogram shows right axis deviation with features of ventricular inversion.

Cardiac catheterization findings were non-specific. Angiocardiograms, however, may be diagnostic as shown by Mehrizi *et al.* (1966) as well as Liberthson *et al.* (1971). The diagnostic method of choice in cases with noninverted ventricles appears to be injection of contrast material into the right atrium yielding simultaneous opacification of both ventricles. For those cases with inverted ventricles, injection of contrast into the left atrium would be expected to yield similar results. In one of our cases, the laevophase of pulmonary arteriography had shown such changes but was misinterpreted.

Another feature based on anatomical findings deserves mention. A right atriogram appears to be the ideal method with which to diagnose double inlet left ventricle in the presence of noninverted ventricles. If a left ventriculogram is done, it may opacify the right atrium. While such an observation brings up for consideration a left ventricular-

right atrial communication, it should be borne in mind that if the tricuspid valve were incompetent, a similar picture could be obtained in double inlet left ventricle with noninverted ventricles.

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