Anatomically corrected malposition of the great arteries

Report of 2 cases, one with congenital asplenia; frequent association with juxtaposition of atrial appendages

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Anatomically corrected malpositions are uncommon cardiac anomalies in which the pulmonary artery originates above the morphological right ventricle, and the aorta above the morphological left ventricle. However, because of interposition of abnormal subaortic conal myocardium, there is absence of aortic valve-mitral valve fibrous continuity. The relation between the pulmonary artery and aorta is also abnormal because of the abnormal conal morphology.

Two cases of anatomically corrected malposition of the great arteries are presented, one of which is the first reported in a patient with the congenital asplenia syndrome; in the other, there was left juxtaposition of the atrial appendages. Review of all known cases of anatomically corrected malposition reveals a significant association with juxtaposition of the atrial appendages and hence with hypoplasia of the right ventricle, subpulmonary and subaortic obstruction. Possible surgical implications of this association are discussed.

Although Harris and Farber first suggested the concept of anatomically corrected transposition of the great arteries in 1939, recognition and documentation of this unusual entity was first provided by Van Praagh and Van Praagh in 1967.

In anatomically corrected transposition of the great arteries, the aorta arises above the morphological left ventricle but with aortic-mitral discontinuity because of abnormal subaortic conus, and the pulmonary artery originates above the morphological right ventricle. Because of the abnormal conal morphology, abnormal relation between the aorta and atrioventricular valve, and abnormal relation between the great arteries, this entity was initially thought to be a form of transposition (Van Praagh and Van Praagh, 1967). However, because transposition of the great arteries is not literally present (the aorta does not originate above the morphological right ventricle and the pulmonary artery does not arise above the left

ventricle), more correctly, then, a positional anomaly of the great arteries, i.e. a malposition, is present; thus anatomically corrected transposition of the great arteries is more properly designated anatomically corrected malposition of the great arteries (Van Praagh *et al.*, 1971).

It is the purpose of this communication to present two further cases of anatomically corrected malposition of the great arteries (one documented at necropsy and the other by angiocardiology) and the first case documented in a patient with the congenital asplenia syndrome, to review other known cases, and to comment on the frequent association with juxtaposition of the atrial appendages.

Case reports

Case 1

This male infant was admitted to the Children's Hospital Medical Center, Boston, Massachusetts, at 7 weeks of age for evaluation of cyanotic congenital heart disease and bloody diarrhoea.

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FIG. 1 Case 1. Admission PA chest radiograph shows dextrocardia, slightly diminished pulmonary vascularity, and abnormally symmetrical hepatic shadow.

Physical examination revealed a cyanosed, obtunded, dehydrated infant with abdominal distension. Dextrocardia was diagnosed; a soft systolic murmur and single second sound were heard. Bowel sounds were absent.

Admission chest radiograph (Fig. 1) showed dextrocardia with an unusual cardiac silhouette and diminished pulmonary vascularity. A symmetrical hepatic silhouette and visceral heterotaxia were evident. A flat plate of the abdomen suggested intestinal obstruction, but a barium enema was unremarkable.

An electrocardiogram revealed a frontal QRS and P axis of $+90^\circ$ and $+120^\circ$, respectively, with a negative P wave in lead I, right atrial enlargement, and right ventricular hypertrophy. T wave inversion was present in the limb and praecordial leads.

Hospital course Shortly after admission the infant sustained a cardiorespiratory arrest requiring intubation and artificial ventilation. After resuscitation, a left pneumothorax was present and was drained with needle thoracostomy. Abdominal distension became more prominent, and mesenteric artery thrombosis secondary to polycythaemia and dehydration was suspected; subsequently, he underwent an exploratory laparotomy where the mid-ileum was found to be oedematous and cyanotic, and the distal ileum was obviously necrotic. The distal ileum was resected and a Mikulicz ileostomy was performed, but the infant died 10 hours after operation before cardiac catheterization could be performed.

Salient findings at necropsy included: dextrocardia; asplenia, abdominal heterotaxy but with basic situs solitus of atria; right superior vena cava and right inferior vena cava draining into the morphological right atrium, right-sided; the left hepatic veins drained separately into the morphologically left atrium, left-sided; left superior vena cava to left atrium, left-sided;

FIG. 2 Case 1. The diminutive right ventricle (RV) is seen. There is a slit-like, subcrystal ventricular septal defect (VSD). The subpulmonary conal myocardium (C) is well seen. The pulmonary artery (PA) originates above this diminutive right ventricular outlet chamber.

small primum atrial septal defect; normal pulmonary venous return to morphologically left atrium, left-sided; a common atrioventricular valve emptying wholly into the morphologically left ventricle, left-sided; a rudimentary outflow chamber, and almost complete absence of the right ventricular sinus, right-sided (D-loop) (Fig. 2); subcrystal ventricular septal defect, slit-like, obstructively small (Fig. 2); crista supraventricularis largely above the morphologically left ventricle (Fig. 3), but also above the very hypoplastic right ventricular sinus; a bilateral conus (subpulmonary and subaortic) preventing fibrous continuity between either semilunar valve and atrioventricular valve; mild infundibular and valvar pulmonary stenosis with a bicuspid pulmonary valve and a defective right septal-left septal commissure, but with severe pulmonary outflow obstruction caused by the small obstructive ventricular septal defect; L-malposition of the aorta with the aorta to the left and slightly anterior (almost side by side) to the pulmonary artery, and originating entirely above the morphologically left ventricle (Fig. 4); a right aortic arch; probe-patent right ductus arteriosus; an abnormally symmetrical liver; pulmonary isomerism with bilateral eparterial bronchial anatomy; left-sided stomach and right-sided gall bladder; abnormal small bowel mesentery; semi-annular pancreas and horseshoe adrenal gland; multiple peripheral emboli with focal renal infarction; and thrombosis of the superior mesenteric

FIG. 3 Case 1. The posterior ventricle, the morphological left ventricle (LV) is seen. This ventricle is moderately hypertrophied. The aorta (AO) is seen to originate above this ventricle. A distinct muscular band, the subaortic conus (C), prevents aortic-mitral fibrous continuity.

FIG. 4 Case 1. The apex of the heart is rotated slightly superiorly to show the relation between the great arteries. The aorta (AO) is to the left of the pulmonary artery (PA). This great vessel relation suggests the 'corrected' transposition relation.

artery with infarction of long segments of the distal jejunum, ileum, and proximal ascending colon.

This infant with the classic features of the asplenia syndrome had a right-sided heart with basically situs solitus of the atria, with a morphologically right atrium, right-sided, receiving the right superior and inferior vena cava. There was atrioventricular concordance as situs solitus of the atria is concordant with a D-loop. The ventricular morphology is typically characteristic of a single left ventricle with a rudimentary outflow chamber and pronounced hypoplasia of the right ventricular sinus. Hence, despite the presence of bilateral conal myocardium, the great vessels were above the morphologically appropriate ventricle. Though the circulation was potentially normal, this was impaired by the common mixing of systemic venous and pulmonary venous blood at the ventricular level.

Case 2

This 14-year-old boy has been followed since infancy for cyanotic congenital heart disease.

Physical examination showed a small, cyanosed boy in no distress. Moderate clubbing was evident. Cardiac examination revealed a prominent left ventricular impulse. The first sound was of normal intensity. The second sound was loud and single at the left upper sternal border. A harsh systolic ejection murmur was best appreciated along the left sternal border. All pulses were full and equal. No hepatomegaly was evident.

The chest radiograph (Fig. 5) showed a peculiar

FIG. 5 Case 2. Standard PA chest radiograph shows abdominal situs solitus. The heart has a flat right heart border and a peculiar silhouette. The heart size is normal, and the pulmonary vascularity appears normal.

cardiac silhouette, with a flat right heart border and diminished pulmonary artery segment. The pulmonary vascularity is slightly diminished. The abdominal situs appears normal. The aortic arch is left sided.

The electrocardiogram reveals left axis deviation of -75° , right atrial enlargement, and left ventricular enlargement.

A peripheral venous angiogram performed by injection of contrast into the right antecubital vein showed opacification of the superior vena cava, right atrium, hepatic veins, and also left juxtaposition of the atrial appendages, and the right ventricular window of tricuspid atresia (Fig. 6). With the catheter positioned in the left ventricle through the atrial septal defect, a left ventricular angiogram was performed. This showed a dilated and hypertrophied morphological left ventricle, left sided, and a hypoplastic right ventricle (Fig. 7 and 8a and b). Hence the ventricles are normally related and are concordant to the atria. The aorta originates solely above the morphological left ventricle, but is abnormally related to its atrioventricular valve. Mitral-aortic fibrous continuity is not present because of interposition of abnormal subaortic conus. The pulmonary valve arises above the hypoplastic right ventricular outflow chamber, and subpulmonary conus results in significant subvalvular pulmonary obstruction. Hence bilateral conus, subaortic and subpulmonary, is present. The aortic valve is abnormally related to the pulmonary valve, the aortic valve being more leftward and anterior than normal. The left pulmonary artery is either absent or severely hypoplastic.

FIG. 6 Case 2. A peripheral venous angiogram from the right median basilic vein shows opacification of the superior vena cava, right atrium, inferior vena cava, and hepatic veins. The right atrial appendage (RAA) is clearly juxtaposed to the left and is superior to the left atrial appendage (LAA). The left ventricle (LV) is clearly opacified. The right ventricular window (W) is outlined.

In summary, this patient had left juxtaposition of the atrial appendages, tricuspid atresia, D-ventricular loop with a hypoplastic right ventricle, subaortic conus preventing mitral-aortic continuity but with the aorta originating solely above the morphological left ventricle and abnormally leftward (L-malposition), subpulmonary conus with significant infundibular and valvar obstruction, ventricular septal defect, atrial septal defect, left aortic arch, and probable absence of the left pulmonary artery.

Discussion

Anatomically corrected transposition of the great arteries is a most uncommon condition and, if transposition is defined literally, then transposition is not present, but a malposition is. The term 'anatomically corrected malposition of the great arteries' has been recently proposed for this entity (Van Praagh *et al.*, 1971).

FIG. 7 Case 2. Frame from selective left ventricular cine angiogram in the antero-posterior projection. The venous catheter has been advanced from the saphenous vein, across the atrial communication into the left ventricle (LV). Contrast material opacifies a large ventricle, clearly a morphological left ventricle, and a diminutive right ventricular outlet chamber (RV). The right pulmonary artery (RPA) is clearly seen. The aorta (AO) is clearly to the left of the pulmonary artery. Bilateral conal myocardium (C) is evident.

In this malposition, the great arteries and ventricles appear to have twisted in opposite directions, resulting in a severe conoventricular malalignment, with a ventricular defect at the conoventricular junction (Van Praagh and Van Praagh, 1967). Bilateral conal myocardium (subaortic and subpulmonary) is usually, but not invariably, present. The abnormal subaortic conal myocardium is always present and separates the aortic and mitral valve. In addition, the subpulmonary conus is frequently present, separating the pulmonary from the mitral valve. Recently, however, Van Praagh has cited an example of anatomically corrected L-malposition of the great arteries with pulmonary-mitral continuity, i.e. only the subaortic conal myocardium is present (Van Praagh, 1973a).

Bilateral or combined conal myocardium is most frequently found in double outlet right ventricle, transposition of the great arteries with subpulmonary stenosis or atresia, the asplenia syndrome, the juxtaposition of atrial appendages syndrome, and usually in anatomically corrected malpositions (Van Praagh, 1973a, c). Goor, Dische, and Lillehei (1972) have recently shown that subaortic conal absorption accounts for aortic-mitral fibrous con-

tinuity. One might speculate, then, that persistence of bilateral conal myocardium in the human represents in part a developmental arrest at 20 to 30 days ovulation age (Horizon 15).

Of 13 adequately documented cases of anatomically corrected transposition (malposition) of the great arteries, 7 had left juxtaposition of the atrial appendages and an additional patient had right juxtaposition of the atrial appendages (Table). Melhuish and Van Praagh (1968) had previously briefly mentioned this association. It is of interest that left juxtaposition of the atrial appendages is normal to about 27 days ovulation age (Horizons 12-13) (Van Praagh, 1973b). Hence, left juxtaposition of the atrial appendages may be interpreted as representing the abnormal persistence of a normal embryonic condition. Significant subvalvar pulmonary obstruction (9/13), tricuspid atresia or hypoplasia (6/13), and hypoplasia of the right ventricular sinus (8/13) are common in anatomically corrected malpositions (Table) and in patients with juxtaposition of the atrial appendages.

Anatomically corrected malpositions had previously been documented in patients with visceral situs solitus (Van Praagh and Van Praagh, 1967;

TABLE Comparative anatomical findings in patients with anatomically corrected malposition of the great arteries

Case No., sex, and age at death	Heart position	Viscero-atrial situs	Ventricular loop	Great vessels	Atrial septum	Atrial appendages	AV valves	Morphological rt. ventricle
1, M, 7 wk	Dextrocardia	Asplenia	D	L-malposition	ASD I	Normal	Common	Hypoplasia of sinus
2, M, 14 yr*	Laevocardia	Solitus	D	L-malposition	ASD II	Left juxtaposition of atrial appendages	Tricuspid atresia	Hypoplasia of sinus
3, F, 2 yr	Dextrocardia	Solitus	D	L-malposition	ASD II, large	Left juxtaposition of atrial appendages	Tricuspid atresia	Hypoplasia of sinus
4, F, 4 wk	Laevocardia	Solitus	D	L-malposition	ASD II, large	Left juxtaposition of atrial appendages	Hypoplasia of tricuspid valve	Hypoplasia of sinus
5, M, 4 wk	Laevocardia	Solitus	L	D-malposition	Intact	Normal	Normal	Hypoplasia of sinus
6, not known, 3 mth	Dextrocardia	Not known	D	L-malposition	PFO	Left juxtaposition of atrial appendages	Normal	Normal
7, F, 7 mth	Dextrocardia	Solitus	D	L-malposition	ASD II	Left juxtaposition of atrial appendages	Tricuspid atresia	Hypoplasia of sinus
8, F, 13 mth	Laevocardia	Solitus	D	L-malposition	ASD II	Left juxtaposition of atrial appendages	Tricuspid atresia	Hypoplasia of sinus
9, F, 1 dy	Mesocardia	Solitus	Malrotated D	D-malposition	ASD II	Right juxtaposition of atrial appendages	Hypoplasia of tricuspid valve	Hypoplasia of sinus
10, F, 4 yr	Laevocardia	Solitus	D	L-malposition	PFO	Normal	Normal	Normal
11, M, 3 mth	Dextrocardia	Inversus	D	L-malposition	Intact	Normal	Normal	Not known
12, M, 31 yr†	Dextrocardia	Solitus	D	L-malposition	Intact	Normal	Normal	Normal
13, F, 8 yr†	Dextrocardia	Solitus	D	L-malposition	Intact	Left juxtaposition of atrial appendages	Normal	Normal

Note: Cases 1 and 2, present cases; Cases 3, 4, and 5, Cases 1, 2, and 3, respectively, of Van Praagh and Van Praagh (1967); Case 6 Raghib (1966); Case 7, Case 16 of Melhuish and Van Praagh, 1968; Case 8 (Stewart and Wynn-Williams, 1956); Case 9, Case 6 of Wagner *et al.*, Case 10 (Sunada *et al.*, 1967); Case 11 (Anderson *et al.* 1972), Cases 12 and 13, Cases 1 and 2 of Kirklin *et al.* (1973).

* Alive. Diagnosed at cardiac catheterization.

† Alive. Diagnosed at cardiac catheterization and confirmed at operation.

Abbreviations: O, absent; D-loop, non-inverted ventricles with morphologically right ventricle, right-sided; L-loop, inverted ventricles; position, malpositioned aortic valve is to the right of pulmonary valve; L-malposition, malpositioned aortic valve is to the left of pulmonary valve; ASD I, ostium primum atrial septal defect; ASD II, secundum type of atrial septal defect; VSD, ventricular septal defect.

Wagner, Alday, and Vlad, 1970) visceral situs inversus (Anderson, Arnold, and Jones, 1972), and now in the congenital asplenia syndrome with visceral heterotaxia. A bilateral conus is most common in the asplenia syndrome (Ivemark, 1955), yet in a necropsy review of 32 patients with congenital asplenia, an anatomically corrected malposition was found in only one case (Case 1) and juxtaposition of the atrial appendages in none (Freedom, 1971; Freedom and Fellows, 1973). This might possibly be explained by the observation that left

juxtaposition of the atrial appendages is normal to 27 days ovulation age, and that the congenital asplenia syndrome results from a slightly later developmental arrest (i.e. after the normal leftward swing of the ventricular apex results in the right atrial appendage passing behind, and ending to the right of the conotruncus).

Although gross pathological evidence of anatomically corrected malposition is lacking in our second case, we feel that this diagnosis is overwhelmingly supported by the angiocardiographic findings. The

Morphologically lt. ventricle	Ventricular septum	Type of conus	Semilunar valve AV valve continuity	Outflow tract obstruction	Aortic arch	Additional anatomical data
Transposition and enlargement	VSD	Bilateral	o	Valvar and infundibular pulm. stenosis	Right	Double inlet left ventricle, right persistent ductus arteriosus
Transposition and enlargement	VSD	Bilateral	o	Valvar and infundibular pulm. stenosis	Left	—
Transposition and enlargement	VSD	Bilateral	o	Valvar and infundibular pulm. stenosis	Right	Probe patent ductus arteriosus
Transposition and enlargement	VSD	Bilateral	o	Infundibular and valvar pulm. atresia	Right	Right persistent ductus arteriosus
Transposition and enlargement	VSD	Bilateral	o	Subaortic stenosis, moderately severe	Left	Hypoplasia of ascending aorta, preductal coarctation
Transposition and enlargement	Multiple VSDs	Bilateral	o	Subaortic stenosis, mild	Left	Mild cor triatriatum, left superior vena cava to coronary sinus
Transposition and enlargement	VSD	Bilateral	o	Valvar and infundibular pulm. stenosis	Right	Absent left coronary ostium
Transposition and enlargement	VSD	Bilateral	o	Infundibular and valvar pulm. atresia	Left	—
Transposition and enlargement	VSD	Bilateral	o	Infundib. pulm. stenosis; subaortic stenosis	Left	Hypoplasia of ascending aorta and arch, left persistent ductus arteriosus
Unknown	VSD	Bilateral	o	None	Left	Persistent ductus arteriosus, left superior vena cava
Normal	VSD	Bilateral	o	None	Not known	Not known
Normal	Multiple VSDs	?Deficient	+	Infundibular pulm. stenosis	Not known	—
Normal	VSD	Bilateral	o	Valvar and infundibular pulm. stenosis	Left	Overriding aorta

aorta originates above the morphologically left ventricle, but there is interposition of the abnormal subaortic conus separating the aortic from the mitral valve; the semilunar valves are abnormally related, the aortic valve being more anterior than normal, and finally the great arteries are not normally related, the aorta being abnormally leftward. This diagnosis has been made only rarely before death, but the angiographic features of this entity seem quite specific (Kirklin *et al.*, 1973). Because the aorta is abnormally leftward and anterior in this malposition, it has been confused angiographically with corrected transposition of the great arteries (Raghib, Anderson, and Edwards, 1966) in which the aorta is abnormally anterior and to the left of the pulmonary artery. In corrected transposition, however, there is both ventricular inversion and transposition, with the aorta originating above the

morphological right ventricle, left-sided (in situs solitus). A selective ventriculogram in the left-sided ventricle will show a morphological right ventricle and the transposed aorta in the patient with corrected transposition of the great arteries. Also, the electrocardiogram and vectorcardiogram will prove useful in this differentiation (Ruttenberg *et al.*, 1966). Kirklin and his associates (1973) have suggested that anatomically corrected malposition of the great arteries might be confused with double outlet left ventricle or double outlet right ventricle with an L-malposed aorta. Selective right and left ventricular biplane ventriculograms will help to distinguish these anomalies from anatomically corrected malpositions.

In addition to the obvious embryological and pathological importance, clinical recognition of anatomically corrected malpositions may have

FIG. 8a and b. Case 2. Selected frames from the left ventricular biplane angiogram in the lateral projection. The semilunar valves are at approximately the same level in this projection. The pulmonary artery (PA) originates above the right ventricular outlet chamber. The aortic valve is clearly separated from the mitral valve (MV) by subaortic conal myocardium (C). This subaortic conal myocardium is quite dynamic. The RV infundibulum (INF) is very narrow and becomes even narrower with systole. Bilateral conal myocardium is evident in this projection.

important surgical implications. Fontan's ingenious operation for the correction of tricuspid atresia (Fontan and Baudet, 1971) uses the right atrial appendage as the proximal end of a valved conduit to the proximal right pulmonary artery. As pointed out earlier, however, and as illustrated by our second case, tricuspid atresia and left juxtaposition of the atrial appendages occur frequently with anatomically corrected malpositions (Table). Hence, correction of tricuspid atresia in the condition using a valved conduit might prove more difficult. It also became apparent that subaortic stenosis occurred not infrequently in this malposition (Table), resulting from the poorly expanded subaortic conal myocardium. If unrecognized, and unrelieved, this condition would seriously jeopardize Fontan's operation, and might possibly be considered a contraindication to it. It is of interest that subaortic obstruction (caused by an obstructively small

ventricular defect) was suggested as the cause of death in a 7-year-old girl with tricuspid atresia, transposition of the great arteries, and severe pulmonary valve and infundibular atresia who underwent Fontan's operation (Ross and Somerville, 1973).

It should also be emphasized that anatomically corrected malpositions may occasionally have a normal tricuspid valve and normal right ventricular size (Raghib *et al.*, 1966; Sunada *et al.*, 1967) and that the intracardiac defects (atrial and ventricular defects) may be amenable to standard intracardiac repair (Sunada *et al.*, 1967). Recently complete cardiac repair with survival was reported in two patients with anatomically corrected malposition of the great arteries (Kirklin *et al.*, 1973). Their first patient, a 31-year-old man, also had subpulmonary obstruction and multiple ventricular septal defects. Left juxtaposition of the atrial appendages, valvar

and subvalvar pulmonary obstruction, ventricular septal defect, and overriding aorta was repaired in their second patient, an 8-year-old girl.

Also, while juxtaposition of the atrial appendages is usually associated with severe congenital cardiac disease, this is not invariably so. Becker and Becker (1970) described right juxtaposition of the atrial appendages in an anencephalic female stillborn; in this infant the great arteries were normally related, and the only intracardiac malformations were an atrial septal defect and a bicuspid pulmonary valve.

Recently bifid right atrial appendage with partial juxtaposition has been described (Charuzi *et al.*, 1973). This pattern was found in 9 of 15 specimens with left-sided juxtaposition and was associated with the same spectrum of complex cardiac malformations commonly associated with complete left-sided juxtaposition.

In summary, anatomically corrected malpositions are rare conotruncal-ventricular disturbances, characterized anatomically by bilateral conus but without transposition of the great arteries. This malposition frequently occurs with juxtaposition of the atrial appendages, tricuspid atresia or hypoplasia, and hypoplasia of the right ventricle. Antemortem recognition of this anomaly is possible because of distinct angiocardiographic features. Awareness of this entity has significant embryological and surgical implications.

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