Approach to Dextrocardia in Adults: *Review*

Pierre D. Maldjian[1] and Muhamed Saric[2]

OBJECTIVE

The educational objectives of this article are to describe an approach to analyzing imaging studies in adults with dextrocardia and to present the appearances of the most common underlying disorders. Topics reviewed include the morphology of the cardiac chambers, the concept of situs, and the relevant embryologic principles. The disorders discussed include situs inversus totalis (mirror-image dextrocardia), dextroversion, congenitally corrected transposition of the great arteries, and polysplenia syndrome.

CONCLUSION

In this article we describe an approach to dextrocardia in adult patients and illustrate the imaging manifestations of the most common underlying disorders.

Introduction

Dextrocardia is a cardiac positional anomaly in which the heart is located in the right hemithorax with its base-to-apex axis directed to the right and caudad. The malposition is intrinsic to the heart and not caused by extracardiac abnormalities. Dextrocardia should be differentiated from cardiac dextroposition, which is defined as displacement of the heart to the right secondary to extracardiac causes such as right lung hypoplasia, right pneumonectomy, or diaphragmatic hernia [1]. Although dextrocardia in infants can be associated with many cardiac anomalies, dextrocardia in adults has a limited range of diagnostic possibilities. As a result of recent advances in CT technology, radiologists are interpreting an increasing number of cardiac imaging studies and will encounter cases of cardiac malposition. Although *situs inversus totalis* is easily recognizable, imagers should also be familiar with the more unusual causes of dextrocardia.

In this article we discuss an approach to dextrocardia in adults and illustrate the appearances of the most common underlying disorders. Before doing so, we set the groundwork by reviewing the morphology of the cardiac chambers, the concept of situs, and the embryologic principles key to understanding these malformations.

Morphology of the Cardiac Chambers

In individuals with normal anatomy, the right cardiac chambers differ in morphology from the corresponding left-sided chambers. Normally, the morphologic right atrium and right ventricle are to the right of the morphologic left atrium and left ventricle. In congenital cardiac malposition, the location of a chamber relative to its morphologic counterpart is variable. For example, the venous atrium (the morphologic right atrium) can be anatomically to the right or to the left of the arterial atrium (the morphologic left atrium). One must refer to chamber morphology rather than relative chamber location to maintain a constant frame of reference.

Certain characteristic features can be used to distinguish the cardiac chambers on imaging studies regardless of their relative locations. The morphologic right atrium is most reliably identified as the chamber that receives the inferior vena cava. The morphologic right atrium also has a triangular appendage with a wide ostium. The morphologic left atrium has a long and tubular appendage with a narrow orifice [2]. Although the superior vena cava usually empties into the morphologic right atrium and the pulmonary veins normally drain to the morphologic left atrium, these venous connections can be variable in the setting of congenital heart disease (i.e., anomalous pulmonary venous return) and are not always dependable for determining chamber morphology [3].

Regarding the ventricles, the morphologic right ventricle has a completely muscular portion of the outflow tract, the infundibulum, which separates the inflow tricuspid atrioventricular valve from the outflow semilunar valve. Trabeculations near the apex and along the interventricular septum are prominent. The moderator band, a trabeculation that extends from the septum to the free wall, is also useful in identifying the morphologic right ventricle. The septal leaflet of the atrioventricular valve is displaced slightly toward the apex relative to the septal leaflet of the inflow valve of the morphologic left ventricle. In individuals with normal anatomy, this results in formation of a short atrioventricular septum between the left ventricle and the right atrium. The morphologic left ventricle lacks an infundibulum, which results in fibrous continuity between the bicuspid inflow valve and the semilunar

outflow valve. The ventricle has a smooth septal surface and contains two well-formed papillary muscles [4].

Situs

The concept of situs refers to the configuration of asymmetric structures within an individual. There are three types of situs: situs solitus (normal), situs inversus (mirror image of normal), and situs ambiguous. Situs applies to the pattern of the viscera as a whole and to each asymmetric viscus itself, such as the lung, liver, spleen, and gastrointestinal tract. Situs also applies to the heart as a whole and to each of the cardiac chambers because each is asymmetric. In situs solitus, the right lung has three lobes and an eparterial bronchus (bronchus above the pulmonary artery), whereas the left lung has two lobes with a hyparterial bronchus (bronchus below the pulmonary artery). The larger lobe of the liver is on the right, and the stomach and spleen are on the left. The morphologic left atrium is to the left of the morphologic right atrium. With situs inversus, the left lung has three lobes with an eparterial bronchus and the right lung has two lobes with a hyparterial bronchus, the larger lobe of the liver is on the left, the stomach and spleen are on the right side of the body, and the morphologic left atrium is to the right of the morphologic right atrium. With situs solitus and situs inversus, the atrial situs always corresponds to the visceral situs [5].

Situs ambiguous is a third type of situs in which the arrangement of the organs is not as ordered (termed "heterotaxy") and the relationship between the atria and the viscera is inconsistent. Situs ambiguous tends toward symmetric morphology of normally asymmetric structures (termed "isomerism"). The two disorders associated with situs ambiguous are asplenia syndrome and polysplenia syndrome. Asplenia syndrome is characterized by bilateral right-sidedness (right isomerism), whereas bilateral left-sidedness (left isomerism) is the hallmark of polysplenia syndrome [6]. Typically, in asplenia syndrome (also called Ivemark's syndrome), the spleen is absent and right-sided structures are duplicated, with bilateral trilobed lungs containing eparterial bronchi. Both atria resemble morphologic right atria. The liver is symmetric and horizontal, and the stomach tends to be near the midline. Associated cardiac malformations include common atrioventricular canal, univentricular heart, transposition of the great arteries, and total anomalous pulmonary venous return (seen in most cases). The complex cyanotic cardiac anomalies and abnormal immune status (because of absence of the spleen) result in a poor prognosis, with a mortality rate of up to 80% in the first year [7].

Polysplenia syndrome is characterized by multiple small spleens, usually adjacent to the stomach, with duplication of left-sided structures, especially bilateral bilobed lungs with hyparterial bronchi. Both atria may resemble morphologic left atria. The tendency to symmetry of the abdominal organs is not as pronounced as in asplenia. Cardiac anomalies are not as complex as in asplenic patients. The most common associated cardiovascular anomalies are azygos continuation of the inferior vena cava, partial anomalous pulmonary venous return, atrial septal defect, and endocardial cushion defect. Up to 25% of patients have minor cardiac abnormalities, and the disorder may not be diagnosed until adulthood [8].

Embryologic Considerations

Some knowledge of the embryology of heart formation facilitates comprehension of the disorders underlying cardiac malposition. The human fetal heart develops from a primitive cardiac tube, with the sinus venosus, atrium, ventricle, bulbus cordis, and truncus arteriosus connected in series. The venous and arterial ends are relatively fixed, and growth of the bulboventricular region results in bending of the cardiac tube, forming the morphologically distinct right and left ventricles (Fig. 1). The atria and venous return from the body form simultaneously so the atria are fixed in position early in development by the entering veins. The atrial situs is not affected by the shape of the bulboventricular loop or the final locations of the ventricles [9]. Thus, the atrial situs corresponds to the visceral situs, and abnormalities of atrial position are usually associated with abnormalities in the situs of other organs [7].

The primitive heart tube may loop to the right (dextro- or D-) or to the left (levo- or L-), forming either a D-loop or an L-loop, respectively. The morphologic right ventricle develops from the bulbus cordis, and the morphologic left ventricle develops from the ventricle of the bulboventricular loop, so the direction of the initial formation of the cardiac loop determines the relative locations of the ventricles. Thus, in formation of a D-loop, the morphologic right ventricle is to the right of the morphologic left ventricle. Conversely, in formation of an L-loop, the morphologic right ventricle is to the left of the morphologic left ventricle. The D-loop is the normal (solitus) cardiac loop and the L-loop is a mirror-image (inversus) loop [9]. The direction of the cardiac loop also determines the course of the coronary arteries. With a D-loop, the anterior descending coronary artery is supplied from the left coronary artery. With formation of an L-loop, the coronary arteries are inverted relative to their position in the normal D-loop, and the anterior descending coronary artery is supplied by the right coronary artery. These relationships exist regardless of the presence or absence of transposition of the great arteries (TGA) [10].

In a D-bulboventricular loop, the morphologic right ventricle (bulbus cordis) is situated adjacent to and forms a connection with the right-sided atrium, regardless of whether that atrium is a morphologic right atrium (as in situs solitus) or a morphologic left atrium (as in situs inversus). Similarly, the morphologic left ventricle connects to the left-sided atrium, again regardless of whether the left-sided atrium is a morphologic left atrium (situs solitus) or a morphologic right atrium (situs inversus). In an L-bulboventricular loop, the morphologic right ventricle (which is now anatomically left-sided) connects to the left-sided atrium and the morphologic left ventricle (which is now right-sided) connects to the right-sided atrium—again, regardless of atrial si-

tus. Thus, in situs solitus with formation of an L-bulboventricular loop (instead of the normal D-bulboventricular loop), the right-sided morphologic right atrium connects to a right-sided morphologic left ventricle, and the left-sided morphologic left atrium drains to a left-sided morphologic right ventricle. In situs inversus with formation of a D-bulboventricular loop (instead of the expected L-bulboventricular loop), the left-sided morphologic right atrium drains to a left-sided morphologic left ventricle, and the right-sided morphologic left atrium connects to a right-sided morphologic right ventricle. This is the embryologic basis for the discordant atrioventricular connections seen in congenitally corrected TGA [11].

The type of cardiac loop can also be characterized as concordant or discordant relative to the type of visceroatrial situs. Concordant cardiac loops are the D-loop with situs solitus and the L-loop with situs inversus. Discordant cardiac loops are L-loop with situs solitus and D-loop with situs inversus. With situs ambiguous, as in asplenia or polysplenia, because the visceroatrial situs cannot be determined, the concordance or discordance of the cardiac loop is also not definable [12]. We will return to the significance of concordant and discordant cardiac loops later in our discussion of TGA.

In the early stages of fetal life with situs solitus and formation of a D-bulboventricular loop, the apex of the heart is in the right hemithorax. At the end of the first month of fetal life, the apex of the heart migrates from the right thorax to the left. Conversely, in situs inversus with formation of an L-loop, the apex of the heart swings from the left thorax to the right. In general, regardless of atrial situs, all D-bulboventricular loops should end development with the heart in the left hemithorax (levocardia), and all L-bulboventricular loops should become dextrocardias. Failure of this shift of the ventricular apex can result in dextrocardia with situs solitus (termed "dextroversion") or levocardia with situs inversus (termed "levoversion"). Although the ventricles may be in an abnormal location, the atria and abdominal viscera maintain their situs solitus or situs inversus position. Dextrocardia can result from a D-bulboventricular loop that fails to undergo a shift into the left hemithorax, or from an L-bulboventricular loop that completes its apical shift into the right hemithorax. Failure of rotation of the ventricular apex is associated with discordant cardiac loops. The shift of the ventricular apex may also be incomplete, resulting in mesocardia (midline cardiac apex). Mesocardia is also associated with discordant bulboventricular loops and the heterotaxy syndromes [13].

In the fifth week of embryogenesis, partitioning of the bulbus cordis and the truncus arteriosus occurs. The developing great arteries at the cephalic end of the truncus arteriosus are relatively fixed in position by the embryonic aortic arches. The sixth aortic arch, which becomes the main pulmonary artery, is posterior to the fourth aortic arch, which becomes the aortic arch proper. Growth of the muscular region, the conus, below the semilunar valve of the pulmonary artery moves the root of the pulmonary artery forward to form continuity with the outflow tract of the right ventricle. Growth of the conus also helps to form the infundibulum, separating the inflow and outflow regions of the right ventricle. With proper conal development, the pulmonary trunk twists around the ascending aorta as the now-anterior proximal right pulmonary artery arises from the anterior ventricle (morphologic right ventricle) and leads to the posterior sixth aortic arch. The aorta arises from the posterior ventricle (morphologic left ventricle), forms in continuity with the developing mitral valve, and leads to the more anterior fourth aortic arch. Thus, the proximal great arteries normally cross when viewed from the side [14–16].

Aberrant conotruncal development can result in formation of an infundibulum below the semilunar valve of the ascending aorta, moving it forward to form continuity with the morphologic right ventricle, while the pulmonary artery forms a connection to the morphologic left ventricle. In most of these cases, the main pulmonary artery is posteriorly displaced and oriented parallel to the aorta when viewed from the side, rather than spiraling around the ascending aorta. This disorder is termed "transposition of the great arteries." It is best to think of TGA simply as formation of discordant ventriculoarterial connections rather than overemphasizing the anteroposterior relationship between the aorta and pulmonary artery [17]. Although in most cases of TGA, the aorta is anterior to the pulmonary artery, there are rare exceptions [15, 16]. Van Praagh [5] recommends that aberrations in conotruncal development be regarded as malpositions of the great arteries. Transposition is simply one variety of malposition. Other types of malposition include double-outlet right ventricle, double-outlet left ventricle, and anatomically corrected malposition [5]. These more complicated malformations are beyond the scope of our discussion.

Regarding the relationships between the great arteries at the level of the semilunar valves, for our purposes we need consider only two types: normal and transposition (Fig. 1). In a D-bulboventricular loop with normal development, the pulmonic valve is located anteriorly, superiorly, and to the left of the aortic valve. In the L-bulboventricular loop, the normal relationship between the great arteries is the mirror image (inverted) of the normal (solitus) relationship found in the D-loop. Thus, the aorta arises from a right-sided morphologic left ventricle and the pulmonary artery arises from a left-sided morphologic right ventricle. In this configuration, the pulmonic valve is located anteriorly, superiorly, and to right of the aortic valve. When transposition occurs in the D-bulboventricular loop, the transposed aortic valve is usually located to the right of the transposed pulmonic valve. Hence, this is designated as D-TGA. When transposition occurs in the L-bulboventricular loop, the transposed aortic valve usually lies to the left of the transposed pulmonic valve. Therefore, this is designated as L-TGA [5].

We can now discuss the importance of determining the concordance or discordance of the cardiac loop relative to the atrial situs. Concordant cardiac loops may have either normal relationship of the great arteries or TGA, whereas discordant cardiac loops usually have "corrected" TGA. "Uncorrected" or
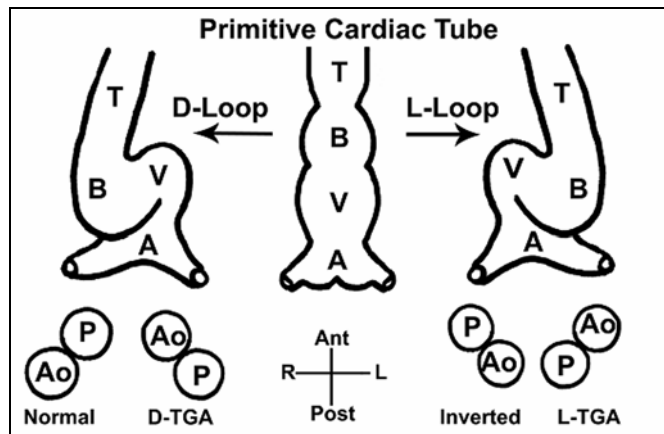
Fig. 1—Diagram illustrates bending of primitive cardiac tube (*top row*) and possible relationships of great arteries at semilunar valve level (*bottom row*). Cardiac tube is depicted from anterior view. Cardiac tube comprises atrium (A), ventricle (V), bulbus cordis (B), and truncus arteriosus (T). Cardiac tube normally bends to right, forming a D-bulboventricular loop. Rarely, tube may bend leftward, forming L-bulboventricular loop. Possible relationships of great vessels are depicted axially from inferior perspective (as seen on CT images in transverse plane). In D-bulboventricular loop, possible relationships are normal or D-TGA (transposition of great arteries). With L-bulboventricular loop, possible relationships are inverted, or L-TGA. Ao = aorta, P = pulmonary artery.

"complete" TGA represents a concordant cardiac loop (D- or L-) with TGA. In a concordant cardiac loop with TGA, the morphologic right atrium empties into a morphologic right ventricle that is connected to the aorta, and the morphologic left atrium empties into a morphologic left ventricle that is connected to the pulmonary artery. Hence, the pulmonary and systemic circulations are independent parallel circuits requiring bidirectional intracardiac or extracardiac shunts to sustain life. Congenitally "corrected" TGA is a discordant cardiac loop. The transposition is said to be corrected because the morphologic left atrium empties into a morphologic right ventricle that connects to the aorta, and the morphologic right atrium empties into a morphologic left ventricle that connects with the pulmonary artery. The discordant atrioventricular connections (a manifestation of the discordant cardiac loop) combine with the discordant ventriculoarterial connections (the definition of TGA) to maintain separation of the systemic and pulmonary circulations.

The classic type of corrected TGA is a discordant cardiac L-loop: situs solitus with L-loop ventricles and L-TGA. The rare form of corrected TGA (because situs inversus is rare) is a discordant D-loop: situs inversus with D-loop ventricles and D-TGA. Note that corrected TGA with situs solitus is L-TGA while corrected TGA with situs inversus is D-TGA. Both of these forms of corrected transposition can be associated with dextrocardia [9]. It is unclear why discordant cardiac loops have such a strong association with TGA, resulting in congenitally corrected TGA. The discordant cardiac loop must somehow influence conotruncal development favoring ventriculoarterial discordance.

Approach to Analysis

The radiologic analysis of dextrocardia (or for that matter, of any congenital heart disease) requires a systematic approach. We recommend the segmental approach, as refined by Van Praagh [5], which proceeds in a logical venoarterial sequence from atria to ventricles to great arteries. Analysis of the following should be performed in sequence: visceroatrial situs, atrioventricular connections, ventricular morphology, ventricular situs, chamber positions, ventriculoarterial connections, and relation of the great

arteries. Finally, any associated anomalies, such as septal defects or pulmonic stenosis, should be described [5].

Because situs of the viscera and that of the atria are almost always the same, the atrial situs is easily seen on cross-sectional imaging. It is also usually readily discernible from the chest radiograph by the location of the liver, spleen, and stomach, as previously described. The morphology of the bronchial tree, if ascertainable, is more accurate than the position of the abdominal organs in determining atrial situs. If there is concern for isomerism, an enlarged azygos vein on chest radiography can be a clue to the presence of polysplenia because azygos or hemiazygos continuation of the inferior vena cava occurs in most cases [18].

Analysis of ventricular morphology, the atrioventricular connections, and the great artery relations usually requires cross-sectional imaging. Important relationships that must be determined are ventricular situs or type of cardiac loop (D- or L-loop), status of the great arteries, and the bulboventriculovisceral relationship (concordant vs discordant cardiac loop) [5]. Using the characteristics previously described, one can differentiate the morphologic right ventricle from the morphologic left ventricle. Van Praagh [5] has described a "hand rule" that can readily determine the ventricular situs. The ventricular situs (D-loop or L-loop) can be thought of as the chirality, or "handedness," and can be decided independently of the location of the ventricle. This is especially useful when the ventricles in a malformed heart show a superoinferior rather than a left-to-right relationship. A D-loop morphologic right ventricle can be considered as right-handed and is present when the palm of the right hand can be placed on the septum, the thumb placed through the inflow, and the fingers placed through the outflow. Similarly, an L-loop morphologic right ventricle can be thought of as left-handed. Conversely, the D-loop morphologic left ventricle is left-handed and the L-loop morphologic left ventricle is right-handed [5, 19]. After a little practice with this method, once having determined the morphology of a ventricle on CT, one can easily decide its situs (D-loop or L-loop) regardless of the position of the heart in the thorax. The considerations pertinent to determining the relationship between the great arteries (normal, inverted, D-TGA, or L-TGA)

Fig. 2—CT scan in 22-year-old man with Kartagener's syndrome. Image of lungs (*left image*) shows dextrocardia and bronchiectasis. Image of great vessels (*right image*) shows that main pulmonary artery (P) is to right of ascending aorta (A), an inverted relationship, as is expected with situs inversus.
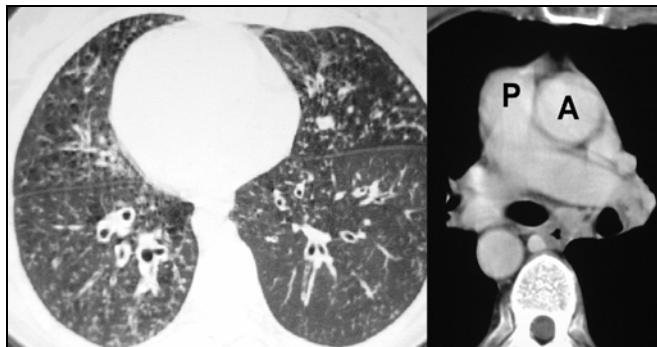
and the concordance or discordance of the cardiac loop have already been discussed.

Disorders Associated with Dextrocardia in Adults

After ascertaining the relationships between the cardiac segments, one can categorize the congenital cardiac anomalies that may be encountered in association with dextrocardia. Theoretically, there are many types of dextrocardia. With situs solitus, one may have dextrocardia with normally related great arteries and D-transposition (complete transposition) or L-transposition (congenitally corrected transposition) of the great arteries. With situs inversus, one may have dextrocardia with inversely related great arteries and D-transposition (congenitally corrected transposition) or L-transposition ("uncorrected" transposition) of the great arteries. With situs ambiguous (either polysplenia or asplenia), one may have dextrocardia with any of the preceding relationships between the ventricles and great vessels. Dextrocardia can also be associated with more complex cardiac malformations, such as single ventricle, double-outlet or double-inlet ventricles, and tricuspid atresia [20].

Because we are concerned with dextrocardia in adults, we need consider only a limited number of possibilities. The most common configuration is dextrocardia with L-loop ventricles and inverted great vessels (situs inversus totalis or the mirror image of normal). Other forms of dextrocardia that may be discovered in adults are situs solitus with D-loop ventricles and normally related great arteries (a variant of dextroversion), situs solitus with L-loop ventricles and L-TGA (congenitally corrected TGA), situs inversus with D-loop ventricles and D-TGA (congenitally corrected TGA), and dextrocardia associated with the polysplenia syndrome.

Dextrocardia with situs inversus, L-loop ventricles, and inverted great arteries results from situs inversus with a concordant L-bulboventricular loop. This is the mirror image of normal and has been called "mirror-image dextrocardia" [1]. Dextrocardia occurs from the migration of the apex of the L-bulboventricular loop into the right hemithorax as expected. This is the most common type of dextrocardia in the general population (present in one or two in 20,000). The incidence of congenital heart disease is low, ranging from 2% to 5% [12].

Kartagener's syndrome occurs in 25% of individuals who have mirror-image dextrocardia (Fig. 2). This disorder is characterized by the triad of situs inversus, paranasal sinusitis, and bronchiectasis. The incidence is estimated to be one or two per 30,000. The primary abnormality is termed "immotile cilia syndrome" or "primary ciliary dyskinesia" and stems from defective ciliary motility due to genetically determined structural abnormalities. Impaired mucociliary clearance leads to recurrent lung infections, bronchiectasis, chronic sinusitis, and otitis media. Ciliary dysfunction also causes reduced fertility in women and sterility in men from reduced motility of spermatozoa [18]. Fifty percent of individuals with primary ciliary dyskinesia have situs inversus. Afzelius [21] has postulated that the beating of primitive cilia might be important in the left–right determination of the early embryo. With ciliary defects, designation of situs in the developing embryo becomes random; hence, half of the individuals have normal situs and half have situs inversus [21].

Dextrocardia with situs solitus, D-loop ventricles, and normally related great arteries results from failure of the final leftward shift of the ventricles during embryologic development. This has been termed "dextroversion" because the heart appears to be rotated into the right hemithorax relative to its normal position. Although the morphologic right atrium and morphologic right ventricle remain to the right, they are located posterior to the corresponding left-sided chambers. It is as if, looking from below, the normal heart is rotated counterclockwise to the patient's right on an axis passing through the right atrium. Actually, the term "dextroversion" is a misnomer because dextrocardia results from failure of rotation into the left hemithorax rather than an abnormal rotation of the apex into the right hemithorax. Dextroversion is the second most common type of dextrocardia. In dextroversion, there is a 90% incidence of additional cardiac malformations, including anomalous pulmonary venous return, tetralogy of Fallot, septal defects, pulmonic stenosis, coarctation of the aorta, and corrected TGA [22]. Isolated dextroversion, without associated congenital cardiac deformities, is rare; but such patients are usually asymptomatic, and discovery of the condition can be delayed until adulthood, when patients present with symptoms of acquired heart disease [23, 24] (Fig. 3).
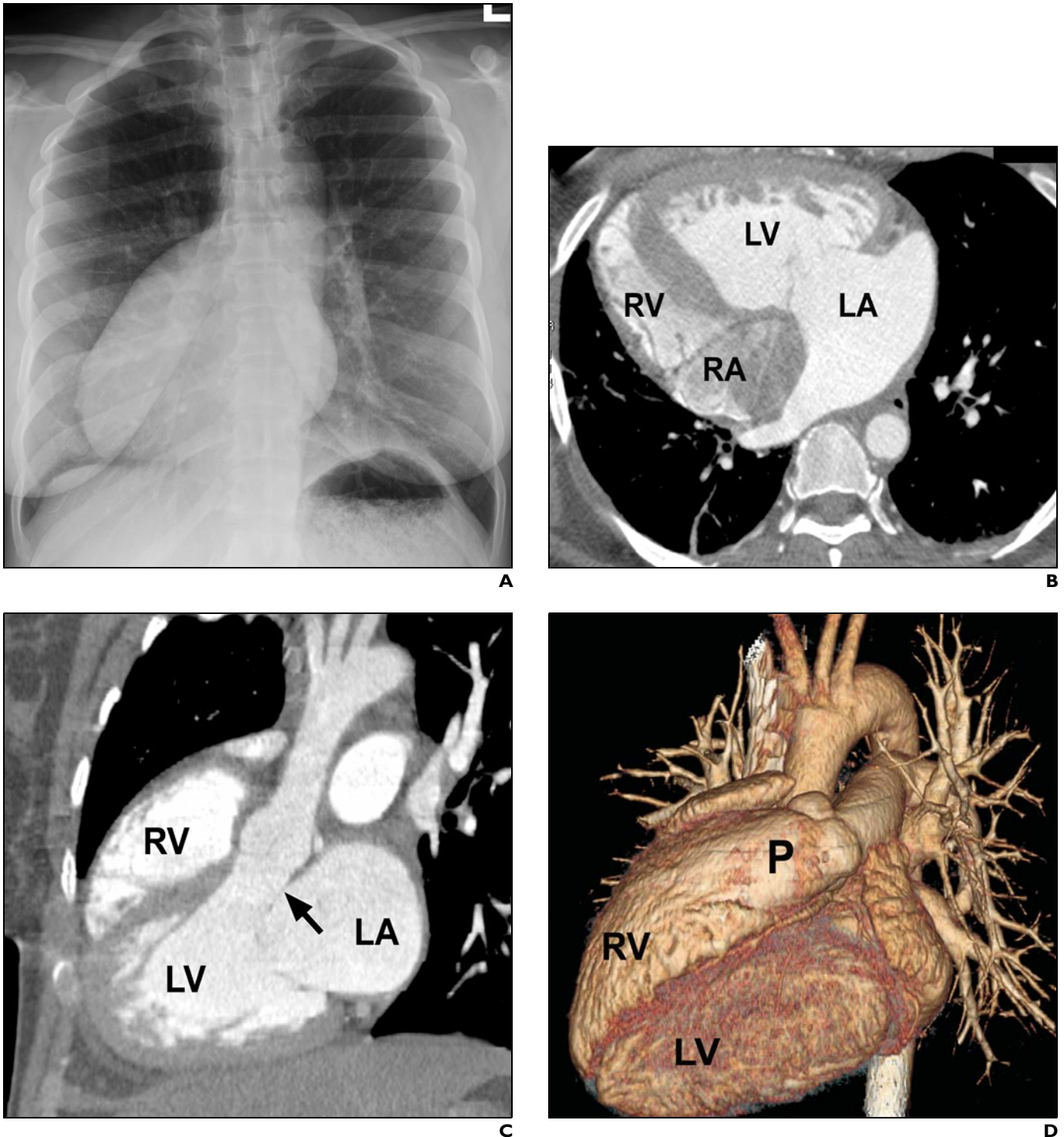
Fig. 3—42-year-old woman with isolated dextroversion.

A, Chest radiograph shows dextrocardia and discordant location of cardiac apex relative to stomach and liver shadow. Locations of gastric bubble and liver shadow are consistent with situs solitus.

B, Axial image from ECG-gated CT scan at level of ventricles shows that left atrium (LA) and left ventricle (LV) are anterior to right atrium (RA) and right ventricle (RV). Ventricles are in D-Loop configuration.

C, Oblique image in plane of outflow tract of left ventricle (LV) shows fibrous continuity between inflow mitral valve (*arrow*) and outflow aortic valve, confirming that left ventricle is a morphologic left ventricle. Left ventricle is located anteriorly and inferiorly to right ventricle (RV).

D, Volume-rendered image in anterior view provides 3D perspective of locations of right ventricle (RV), left ventricle (LV), and pulmonary outflow tract (P) of right ventricle. Muscular outflow tract confirms that right ventricle is a morphologic right ventricle. Pulmonic valve is located anteriorly and slightly to left of aortic root, as in normal patients.

Dextrocardia can also occur in association with congenitally corrected TGA in up to 30% of cases, whereas up to 15% may have mesocardia [12, 25]. Congenitally corrected TGA can occur with situs solitus or, less commonly, situs inversus. Situs solitus with L-loop ventricles is a discordant L-bulboventricular loop, and, with the addition of the associated L-TGA, results in congenitally corrected TGA (Fig. 4). Dextrocardia may occur in these cases from the terminal shift of the ventricular apex during embryology into the right hemithorax (as would be expected in an L-bulboventricular loop).

This form of dextrocardia is also considered a less common variant of dextroversion [26]. (The term "dextroversion" is usually applied in all cases of dextrocardia with situs solitus [17, 27].) However, again the term "dextroversion" is a misnomer because the dextrocardia results from the expected movement of the apex of an L-bulboventricular loop into the right hemithorax rather than an abnormal cardiac rotation. Interestingly, most cases of corrected TGA with situs solitus do not have dextrocardia because the formation of the discordant cardiac loop appears to impede complete migration of
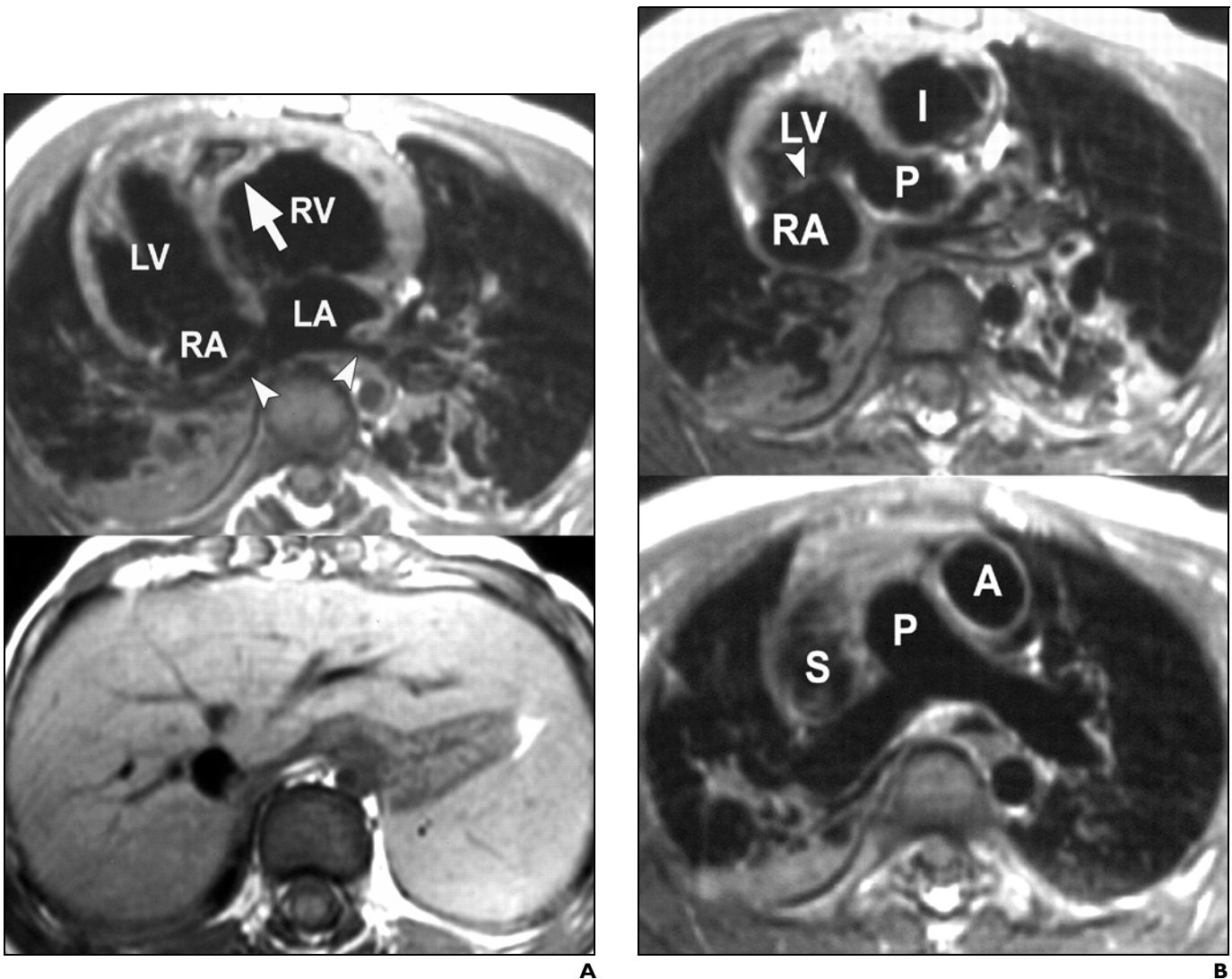

A

B

Fig. 4—ECG-gated axial spin-echo T1-weighted MR images in patient with dextrocardia, situs solitus, and corrected transposition of great arteries (TGA). (Reprinted with permission from Reddy GP, Caputo GR. Diagnosis please: case 15. *Radiology* 1999; 211:709–710 [2])
A, Liver is on right and spleen is on left (*bottom image*), revealing situs solitus. Image through cardiac chambers (*top image*) shows discordant atrioventricular connections. Inferior pulmonary veins (*arrowheads*) drain to morphologic left atrium (LA). Left atrium is connected to morphologic right ventricle (RV), which is distinguished by presence of a moderator band (*arrow*). Morphologic right atrium (RA) is connected to morphologic left ventricle (LV). Ventricles are in L-loop configuration.
B, Images at progressively higher levels show muscular outflow tract or infundibulum (I, *top image*), which is characteristic of a morphologic right ventricle. Pulmonary artery (P) arises from outflow tract of morphologic left ventricle (LV, *top image*). Right-sided atrioventricular valve (*arrowhead, top image*) is near root of pulmonary artery because of fibrous continuity of inflow and outflow valves characteristic of a morphologic left ventricle. In lower image, aortic root (A) arises from morphologic right ventricle. Discordant atrioventricular and ventriculoarterial connections define disorder as congenitally corrected TGA. This case represents L-TGA because aorta is to left of pulmonary artery. RA = right atrium, S = superior vena cava.
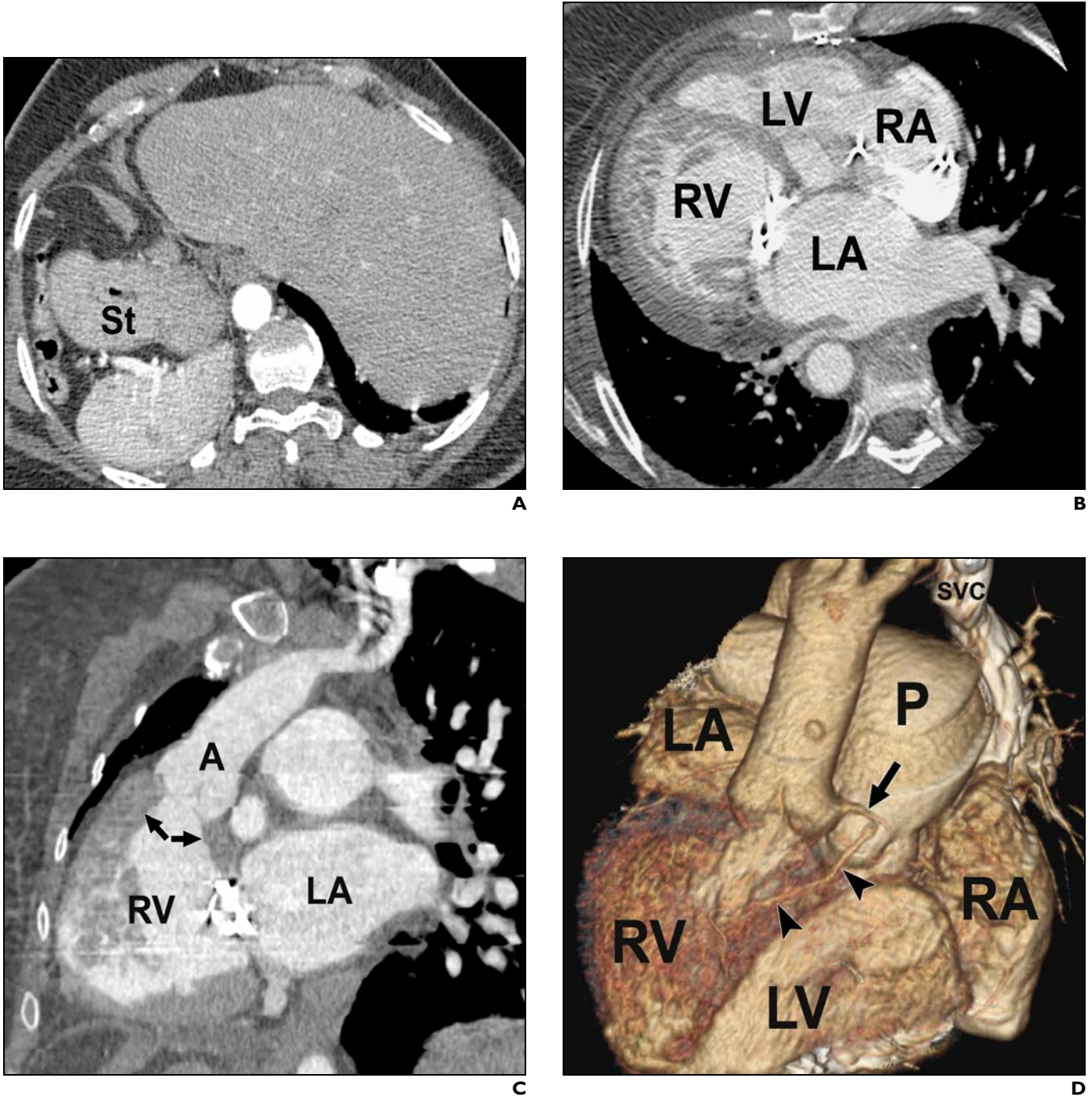
Fig. 5—Images from ECG-gated CT scan of 52-year-old woman with dextrocardia, situs inversus, and congenitally corrected transposition of great arteries (TGA).

A, Axial image through upper abdomen shows liver on left and spleen and stomach (St) on right, consistent with situs inversus.

B, Axial image at level of cardiac chambers shows that morphologic left atrium (LA) is connected to a morphologic right ventricle (RV), distinguished by prominent trabeculations along its septal surface. Morphologic right atrium (RA) is connected to a morphologic left ventricle (LV). Artifacts are caused by presence of pacemaker in left ventricle and prosthetic valve replacement of inflow valve of right ventricle. (Arrhythmias and tricuspid valve dysfunction are common complications of congenitally corrected TGA.) Ventricles are in D-loop configuration.

C, Reformatted oblique coronal image through outflow tract of posterior ventricle shows muscular infundibulum (*arrows*) separating inflow and outflow regions and confirming that posterior ventricle is a morphologic right ventricle (RV). Ventricle connects to aorta (A). LA = morphologic left atrium.

D, Volume-rendered image from anterior view provides 3D perspective of relationships of cardiac chambers and great vessels. Aortic valve is superior and to right of pulmonic valve, as expected with D-TGA. Pulmonary artery (P) is enlarged in this patient due to pulmonary arterial hypertension that is likely secondary to tricuspid valve disorder. Even though patient has situs inversus, anterior descending coronary artery (*arrowheads*) is supplied by left coronary artery (*arrow*) because ventricles are in D-loop configuration. LA = morphologic left atrium, LV = morphologic left ventricle, RA = morphologic right atrium, RV = morphologic right ventricle, SVC = superior vena cava.

the apex of an L-bulboventricular loop into the right hemithorax [25].

Situs inversus, D-loop ventricles, and D-TGA are a discordant D-bulboventricular loop. The D-TGA in conjunction with the discordant D-loop ventricles results in congenitally corrected TGA (Fig. 5). Failure of the terminal shift of the apex of the D-bulboventricular loop into the left hemithorax during embryologic development can result in dextrocardia. This configuration

is rare because situs inversus is rare, and discordant bulboventricular loop development is unusual even in situs inversus [12].

Associated cardiac anomalies, such as ventricular septal defect, pulmonary outflow obstruction, and systemic atrioventricular valve dysfunction, are common in all forms of congenitally corrected TGA [28]. The tricuspid valve often has dysplastic leaflets. Approximately 25% of cases have Ebstein's anomaly of the tricuspid valve without the full spectrum of dysplasia of the
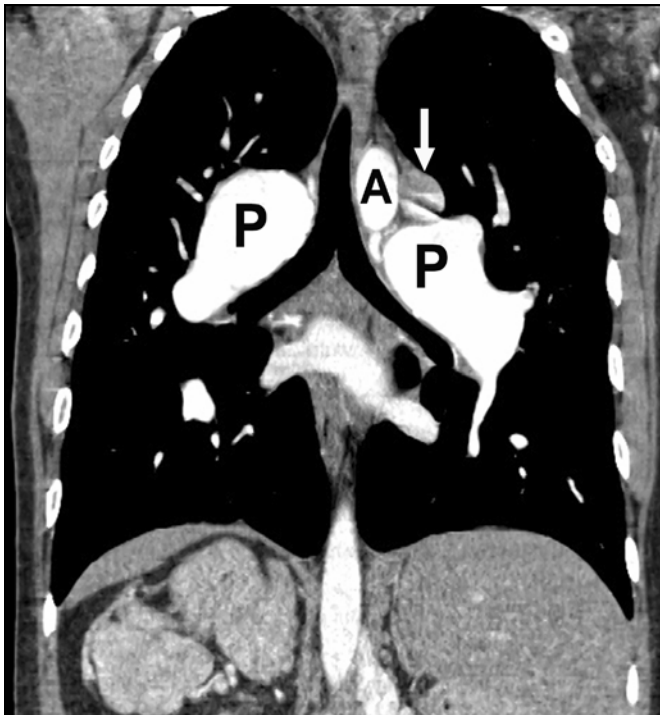
Fig. 6—Images from ECG-gated CT scan of 20-year-old man with dextrocardia and polysplenia syndrome.
A, Axial image through upper abdomen shows multiple splenules in right upper quadrant posterior to right-sided stomach (St). Liver is predominantly left-sided. Vessel (*arrow*) adjacent to descending thoracic aorta is enlarged hemiazygos vein from hemiazygos continuation of inferior vena cava. Note absence of intrahepatic portion of inferior vena cava.
B, Axial image through cardiac chambers shows complete endocardial cushion defect comprised of absence of atrial septum (common atrium) and upper portion of ventricular septum. Arrows indicate leaflets of common atrioventricular valve.
C, Reformatted coronal image shows bilateral hyparterial bronchi. Pulmonary arteries (P) are enlarged from chronic pulmonary artery hypertension secondary to longstanding intracardiac left-to-right shunt. A = aorta, arrow = arch of hemiazygos vein connecting to left-sided superior vena cava (not shown).

morphologic right ventricle that is usually associated with Ebstein's anomaly. Clinically significant tricuspid regurgitation develops in 20–50% of patients. Tricuspid incompetence in these individuals presents in the same way as mitral regurgitation in otherwise healthy patients [29]. A systemic morphologic right ventricle is less tolerant of valvular incompetence than a morphologic left ventricle, and progression of tricuspid regurgitation can lead rapidly to deterioration of ventricular function [30]. Progressive deterioration of function of the morphologic right ventricle with age (beginning from the third decade of life) has also been linked to abnormal perfusion that causes myocardial ischemia or infarction [31].

Arrhythmias, especially complete atrioventricular (AV) block, are also commonly associated with congenitally corrected TGA in up to 30% of patients [2]. The sinoatrial node and atrial portion of the AV node are located in the venous atrium (morphologic right atrium). The AV bundle and bundle branches rotate with the bulboventricular loop. Thus, the course of the conduction system in congenitally corrected TGA is abnormal because the ventricular positions are discordant relative to the morphologic right atrium [32]. Complete AV block may be present at birth, may occur as a complication of surgery from closure of an associated ventricular septal defect, or may occur spontaneously over time. The risk of developing a complete AV block increases linearly with age at the rate of 2% per year [33]. Individuals who develop complete block usually require implantation of a permanent pacemaker. Even when no severe cardiac anomalies are present at birth, the later onset of regurgitation of the systemic atrioventricular valve, dysfunction of the systemic ventricle, or total AV block affects longevity in patients with congenitally corrected TGA, and few patients live past age 50 [29].

Dextrocardia can also be associated with the heterotaxy syndromes of asplenia and polysplenia. Of the two syndromes, polysplenia is more likely to be associated with less severe cardiac malformations, and, therefore, more likely to be encountered in adults (Fig. 6). Up to 50% of cases of polysplenia syndrome can have dextrocardia. The tendency in polysplenia syndrome is toward noncyanotic congenital heart defects [8]. Abnormalities associated with polysplenia syndrome are bilateral symmetric liver (50–67%), bilateral bilobed lungs with bilateral hyparterial bronchi (68–88%), bilateral superior vena cava (33–50%), absence of the intrahepatic portion of the inferior vena cava with azygos or hemiazygos continuation (58–100%), common atrium with complete absence of the atrial septum (25–30%), endocardial cushion defect (80%), hypoplasia or absence of one ventricle (37%), subvalvular or valvular pulmonary stenosis (42–43%), aortic obstruction or atresia (17–22%), and double-outlet right ventricle (17–37%) [34]. Curiously, there also appears to be an unexplained relationship between polysplenia and Kartagener's syndrome. Features of polysplenia syndrome have been reported to occur in patients with Kartagener's syndrome, and primary ciliary dyskinesia has also been reported in patients with the polysplenia syndrome [18].

Summary

In conclusion, we have discussed various disorders that can be encountered with dextrocardia in adults. With situs solitus, one should consider dextroversion of either the D-loop or L-loop variety (the L-loop form represents congenitally corrected TGA). With situs inversus, situs inversus totalis, also called mirror-image dextrocardia, is most likely. However, situs inversus with congenitally corrected TGA, albeit rare, should also be considered. If dextrocardia is associated with isomerism, polysplenia syndrome is the most likely diagnosis. In this article, we have also described a systematic approach for analyzing the cardiac segments. Although the precepts elucidated here are useful for deciphering dextrocardia, they are general principles that can be applied to congenital cardiac disease regardless of the position of the heart in the thorax.

References

1. Bharati S, Lev M. Positional variations of the heart and its component chambers. *Circulation* 1979; 59:886–887
2. Reddy GP, Caputo GR. Diagnosis please: case 15. *Radiology* 1999; 211:709–710
3. Rao S. Dextrocardia: systematic approach to differential diagnosis. *Am Heart J* 1981; 102:389–403
4. Araoz PA, Reddy GP, Higgins CB. Congenital heart disease: morphology and function. In: Higgins CB, De Roos A, eds. *Cardiovascular MRI & MRA.* Philadelphia, PA: Lippincott Williams & Wilkins, 2003:307–338
5. Van Praagh R. The importance of segmental situs in the diagnosis of congenital heart disease. *Semin Roentgenol* 1985; 20:254–271
6. Tonkin IL, Tonkin AK. Visceroatrial situs abnormalities: sonographic and computed tomographic appearance. *AJR* 1982; 138:509–515
7. Applegate KE, Goske MJ, Pierce G, Murphy D. Situs revisited: imaging of the heterotaxy syndrome. *RadioGraphics* 1999; 19:837–852
8. Van Mierop LH, Gessner IH, Schiebler GL. Asplenia and polysplenia syndromes. *Birth Defects: Original Article Series* 1972; 8:36–44
9. Van Praagh R, Van Praagh S, Vlad P, Keith JD. Anatomic types of congenital dextrocardia: diagnostic and embryological implications. *Am J Cardiol* 1964; 13:510–531
10. Van Praagh R. The segmental approach to diagnosis in congenital heart disease. *Birth Defects: Original Article Series* 1972; 8:4–23
11. Shaher RM, Duckworth JW, Khoury GH, Moes CA. The significance of the atrial situs in the diagnosis of positional anomalies of the heart. *Am Heart J* 1967; 73:32–48
12. Ellis K, Fleming RJ, Griffiths SP, Jameson G. New concepts in dextrocardia: angiographic considerations. *AJR* 1966; 97:295–313
13. Elliott LP, Jue KL, Amplatz K. A roentgen classification of cardiac malpositions. *Invest Radiol* 1966; 1:17–28
14. Rosenbaum HD. The roentgen classification and diagnosis of cardiac alignments. *Radiology* 1967; 89:466–476
15. Van Praagh R, Van Praagh S. Isolated ventricular inversion: a consideration of the morphogenesis, definition and diagnosis of nontransposed and transposed great arteries. *Am J Cardiol* 1966; 17:395–406
16. Van Praagh R, Perez-Trevino C, Lopez-Cuellar M, et al. Transposition of the great arteries with posterior aorta, anterior pulmonary artery, subpulmonic conus and fibrous continuity between aortic and atrioventricular valves. *Am J Cardiol* 1971; 28:621–631
17. Wilkinson JL, Cochrane AD, Karl TR. Congenital heart surgery nomenclature and database project: corrected (discordant) transpositions of the great arteries and related malformations. *Ann Thor Surg* 2000; 69:S236–S248
18. Winer-Muram HT. Adult presentation of heterotaxic syndromes and related complexes. *J Thorac Imaging* 1995; 10:43–57
19. Van Praagh S, La Corte M, Fellows KE, et al. Supero-inferior ventricles: anatomic and angiocardiographic findings in 10 postmortem cases. In: Van Praagh R, Takao A, eds. *Etiology and morphogenesis of congenital heart disease.* Mount Kisco, NY: Futura, 1980:317–378

20. Huhta JC, Hagler DJ, Seward JB, Tajik AJ, Julsrud PR, Ritter DG. Two-dimensional echocardiographic assessment of dextrocardia: a segmental approach. *Am J Cardiol* 1982; 50:1351–1360

21. Afzelius BA. Inheritance of randomness. *Med Hypotheses* 1996; 47:23–26

22. Buxton AE, Morganroth J, Josephson ME, Perloff JK, Shelburne JC. Isolated dextroversion of the heart with asymmetric septal hypertrophy. *Am Heart J* 1976; 92:785–790

23. Totaro P, Coletti G, Lettieri C, Pepi P, Minzioni G. Coronary artery bypass grafts in a patient with isolated cardiac dextroversion. *Ital Heart J* 2001; 2:394–396

24. Salanitri JC, Welker M, Pereles FS. Magnetic resonance imaging of acute myocardial infarction in dextrocardia with situs solitus (dextroversion). *Australas Radiol* 2005; 49:422–426

25. Rutledge JM, Nihill MR, Fraser CD, Smith EO, McMahon CJ, Bezold LI. Outcome of 121 patients with congenitally corrected transposition of the great arteries. *Pediatr Cadiol* 2002; 23:137–145

26. Swischuk LD, Sapire DW. *Basic imaging in congenital heart disease,* 3rd ed. Baltimore, MD: Williams & Wilkins, 1986:249–251

27. Gedgaudas E, Miller JH, Castaneda-Zuniga WR, Amplatz K. *Cardiovascular radiology.* Philadelphia, PA: Saunders, 1985:175–178

28. Caso P, Ascione L, Lange A, Palka P, Mininni N, Sutherland G. Diagnostic value of transesophageal echocardiography in the assessment of congenitally corrected transposition of the great arteries in adult patients. *Am Heart J* 1998; 135:43–50

29. Liberthson RR. Congenitally corrected transposition of the great arteries: ventricular inversion or levotransposition. In: Liberthson RR, ed. *Congenital heart disease diagnosis and management in children and adults.* Boston, MA: Little, Brown and Company, 1989:181–186

30. Prieto LR, Hordof AJ, Secic M, Rosenbaum MS, Gersony WM. Progressive tricuspid valve disease in patients with congenitally corrected transposition of the great arteries. *Circulation* 1998; 98:997–1005

31. Hornung TS, Bernard EJ, Celermajer DS, et al. Right ventricular dysfunction in congenitally corrected transposition of the great arteries. *Am J Cardiol* 1999; 84:1116–1119

32. Anderson RH, Becker AE, Arnold R, Wilkinson JL. The conducting tissues in congenitally corrected transposition. *Circulation* 1972; 50:911–923

33. Huhta JC, Maloney JD, Ritter DG, Ilstrup DM, Feldt RH. Complete atrioventricular block in patients with atrioventricular discordance. *Circulation* 1983; 67:1374–1377

34. Bartram U, Wirbelauer J, Speer CP. Heterotaxy syndrome: asplenia and polysplenia as indicators of visceral malposition and complex congenital heart disease. *Biol Neonate* 2005; 88:278–290