Cardiac Malpositions
An Overview Based on Study of Sixty-five Necropsy Specimens

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Cardiac malformations associated with cardiac malpositions are so complex that even angiographic studies may not afford as accurate information as direct examination of necropsy specimens. This communication will attempt to outline a systematic approach to cardiac malpositions based upon necropsy studies of 65 cases. Where possible, morphologic features characteristic of the entities are correlated with clinical, roentgenographic, electrocardiographic and catheterization findings. Embryologic considerations are also discussed.

The problems of terminology in complex congenital cardiac anomalies, particularly those with abnormalities of position, are well known. For an excellent discussion, the reader is referred to the recent article by Wilkinson and Acere.1 Rather than creating new terms, the authors selected existing terms which they considered the least ambiguous and least confusing. We have used the selected terms as defined in that article with three exceptions. 1) The asplenia and the polysplenia syndromes are sufficiently distinct to warrant separate categories, rather than grouping them as situs ambiguous. The reasons for this are discussed in the sections dealing with these entities. 2) Transposition herein refers only to transposition of the great arteries as defined by Van Praagh,2 i.e., aorta arising from the morphologic right, and pulmonary artery from the morphologic left, ventricle. 3) We concur with Van Praagh in classifying Taussig-Bing anomaly as a form of double outlet right ventricle with a subpulmonic ventricular septal defect.8

Definitions

Cardiac malposition. Normally the major portion of the heart lies to the left of midline. For the purposes of this study, any heart other than a left-sided heart in a situs solitus individual represents a cardiac malposition. This definition refers not only to the cardiac position but also the appropriateness of the cardiac position in relation to the total body situs. A right-sided heart is clearly unusual even though it may be appropriate in an individual with situs inversus. A left-sided heart, however, is inappropriate in situs inversus and is also an example of cardiac malposition.

For theoretical reasons to be discussed later, all cases of asplenia and polysplenia are regarded as having cardiac malpositions.

Levocardia, dextrocardia and mesocardia are general terms indicating cardiac position only. They do not give any indication of cardiac structure, body situs, or electrocardiographic findings. Levocardia denotes a left-sided heart; dextrocardia denotes a right-sided heart; and mesocardia denotes a midline heart.

Cardiac displacement is the shifting of the heart within the thorax by extracardiac factors such as a hypoplastic lung. This has also been referred to in the literature as dextroposition, mesoposition or levoposition, depending on the direction of the shift.

Inversion is an alteration in the lateral relationships of asymmetric body structures so that a structure which normally lies on the right side of the body is situated on the left and vice versa. Inversion may involve the entire body (situs inversus) or only certain structures such as the great arteries.

Heterotaxy is abnormal arrangement of organs different from the orderly arrangement of either situs solitus or situs inversus, e.g., malrotation of bowel.

Transposition means that the aorta arises from the morphologic right ventricle (RV), and the pulmonary artery from the morphologic left ventricle (LV). With rare exceptions,3 there is aortic-mitral discontinuity. The aorta is usually anterior to, but may be lateral to, the main pulmonary artery.

Single LV or single RV is a situation in which one ventricle receives the entire portion of both atrioventricular valves or a common atrioventricular valve. This also has been termed double inlet left ventricle or double inlet right ventricle.

Double outlet right ventricle (DORV) denotes both great arteries arising from the morphologic right ventricle. Usually neither semilunar valve has continuity with the mitral valve (subaortic and subpulmonic conus).

Double outlet left ventricle (DOLV) denotes both great arteries arising from the morphologic left ventricle. The conus anatomy in this situation is variable but is usually bilaterally deficient.

It should be noted that as used herein DORV differs from the broader use proposed by Lev,4 i.e., aorta and pulmonary artery completely or almost completely arising from the right ventricle regardless of semilunar valve mitral continuity. The latter definition focuses entirely on the position of the great arteries. As such it includes cases of tetralogy with severe overriding of the aorta and excludes those cases of Taussig-Bing anomaly in which a substantial portion of the pulmonary artery overrides the left ventricle.6

General Features of Cardiac Anatomy

In describing an anatomic cardiac complex, three segments must be considered: 1) the total body configuration or situs, including the atria; 2) the ventricular positions

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and connections to the atria; 3) the positions of the great arteries and connections to the ventricles.

**Anatomic Features of Body Configuration or Situs**

Although many paired structures of the body are normally symmetrical (e.g., brain, kidneys, ureters), several paired viscera are asymmetric (e.g., tracheobronchial tree, lungs, atria). The liver, although a solitary organ, has two unequal lobes. Solitary structures, such as the spleen and gastrointestinal tract, have a specific rightward-leftward orientation within the abdominal cavity. The positions of these asymmetric structures express the situs or body configuration of an individual. Classically, these have been divided into two asymmetrical body configurations: 1) situs solitus or conventional normal, and 2) situs inversus or complete inversion (fig. 1). In situs solitus, the right lung has three lobes and an eparterial bronchus while the left has two lobes and a hyparterial bronchus. The larger lobe of the liver is on the right and the stomach and spleen are on the left. The morphologic left atrium is posterior and leftward. In situs inversus, the left lung has three lobes and the right lung has two. The tracheobronchial tree and atria are similarly inverted. The larger lobe of the liver is on the left while the stomach and spleen are situated on the right.

In addition, two symmetrical body configurations have been found to be associated with splenic anomalies. *Asplenia* syndrome is characterized by bilateral right-sidedness or duplication of right-sided structures⁸⁻¹⁰ including bilateral right lungs with bilateral eparterial bronchi. Both atria morphologically resemble right atria, the liver is symmetrical and horizontal and the stomach tends to be near the midline. *Polysplenia* syndrome is characterized by a tendency toward bilateral left-sidedness¹¹,¹² which may include bilateral left atria and bilateral left lungs with bilateral hyparterial bronchi. The latter is the most constant feature of symmetry.¹² The abdominal organs tend less toward symmetry than in asplenia. Although the liver may have two roughly equal halves, the major portion often lies to one side of the abdomen. Similarly, the stomach is seldom midline and considerable abdominal heterotaxy is often present. The multiple spleens in this condition are situated adjacent to the stomach and usually resemble a cluster of grapes or a bilobed or tri-lobed spleen. In a given case, the size of the individual spleens may vary but each is much smaller than a normal spleen. This is in contrast to *accessory spleens* wherein one or more spleniculi are present in addition to a normal-sized spleen. The absence of a spleen or the presence of multiple spleens also may be regarded as additional manifestations of the bilateral right-sidedness or left-sidedness, respectively.¹² Although there may be some overlap of the anatomic features of asplenia and polysplenia syndromes, the differences are considerably more frequent and are sufficiently consistent to warrant designations as separate body configurations.

Cases of isolated absence of the spleen, i.e., without associated visceral and cardiac anomalies, are herein regarded as not having the developmental complex of asplenia syndrome.

**Anatomic Features of Cardiac Chambers**

The morphologic features of right and left atria are listed in table 1. Van Praagh has stated that the atria follow the body situs; however, he designated the atria and situs of asplenia as uncertain.¹³,¹⁴ In our experience, the morphologic atria conform to the situs even in cases of asplenia or polysplenia, i.e., bilateral morphologic right atria in asplenia and a distinct tendency toward bilateral left atria in polysplenia. The atrial symmetry has also been reported by Van Mierop.⁶,¹²

The morphologic features of right and left ventricles are listed in table 1 and illustrated in figure 2. When the morphologic right ventricle is situated on the right and the morphologic left ventricle is situated on the left, the ventricles are considered to be noninverted and the result of a d-bulboventricular loop.¹⁵ Conversely, when the morphologic right ventricle is on the left and the morphologic left ventricle is on the right, the ventricles are considered to be inverted and the result of an l-bulboventricular loop.¹⁶ Regardless of where the morphologic right ventricle lies, the infundibulum is almost always the most anterior cardiac structure and connects with the anterior great artery. The left ventricular outflow tract lies posterior to the infundibulum and connects with the posterior great artery. The terms noninversion and inversion of the ventricles correspond to the d- and l-loops in the nomenclature of Van Praagh.¹⁶

The ativoventricular valve of each ventricle is characteristic of that ventricle. While the ativoventricular node and the initial segment of the bundle of His lie in the right atrial wall, the branches of the bundle follow the respective ventricles.¹⁵,¹⁶

A d-loop is normal for situs solitus and an l-loop for situs inversus. Van Praagh termed these *concordant* loops.
Discordant loops are an L-loop in situs solitus and a d-loop in situs inversus. With rare exceptions,17 discordant loops are associated with abnormally related great arteries. We have chosen to designate all cases of splenic anomalies as having discordant loops because symmetry of the atria, by definition, precludes connection with the appropriate morphologic ventricles.

Anatomic Features of the Great Arteries

The anatomy of the great arteries may best be defined in terms of their lateral interrelationships and ventricular attachments. Normally attached great arteries are characterized by the pulmonary artery arising from the right ventricle and the aorta from the left ventricle. In this situation, the origin of the pulmonary artery lies anterior to the origin of the aorta. As the infundibulum is longer than the left ventricular outflow tract, the pulmonary valve is usually more cephalad than the aortic. In transposition of the great arteries, the origin of the aorta usually lies anterior to that of the pulmonary artery and arises from the infundibulum. As a result, the aorta is the more cephalad semilunar valve and there is no fibrous continuity between the aortic and mitral valves. The latter was considered by some authors an essential feature of transposition;18 however, exceptions have been found.2

The lateral interrelationships of the great arteries are best described using "d" and "l" terms. With "d" related great arteries, the ascending aorta sweeps toward the right and lies to the right of the main pulmonary artery. With "l" related great arteries, the ascending aorta sweeps to the left and lies to the left of the main pulmonary artery. These lateral relationships apply to normally related great arteries, transposed great arteries, and to situations in which both great arteries arise from one ventricle. In addition, there are transpositions in which the aorta lies directly in front of the pulmonary artery, a situation which has been designated as a-trans (for anterotransposition).19

Nomenclature

The original Van Praagh nomenclature14, 20 has been particularly useful in describing complex cardiac anatomy and in focusing attention on the embryological development of complex anomalies. Certain exceptions, however, did not conform to the loop rule and made use of the original Van Praagh nomenclature in these cases difficult, e.g., situs solitus with complete transposition but with the great arteries in the l-transposition configuration. The nomenclature was subsequently enlarged so that in situations where the great artery interrelationship might be confused by the designation d- or l-transposition and in double outlet right ventricle, the term d- or l- malposition was used.21, 22

More recently, Van Praagh has introduced a nomenclature modified from the original. The modification is an entirely symbolic representation of the basic cardiac structure, designating the situs, ventricular interrelationships and great artery interrelationships in that sequence.23, 24 Although the symbolic representation may prove quite useful for pediatric cardiologists and cardiac

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**Table 1. Anatomic Features of Cardiac Chambers**

<table>
<thead>
<tr>
<th>Morphologic right</th>
<th>Morphologic left</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atria</td>
<td></td>
</tr>
<tr>
<td>Lies on same side as trilobed lung.</td>
<td>Lies on same side as bilobed lung.</td>
</tr>
<tr>
<td>Receives the inferior vena cava.</td>
<td></td>
</tr>
<tr>
<td>Crista terminalis.</td>
<td></td>
</tr>
<tr>
<td>Pectinate muscles in the atrial appendage.</td>
<td>Interatrial ostium II on the left side of atrial septum.</td>
</tr>
<tr>
<td>Fossa ovalis lies on the right side of the atrial septum.</td>
<td>Mitral valve.</td>
</tr>
<tr>
<td>Ventricles</td>
<td></td>
</tr>
<tr>
<td>Tricuspid valve.</td>
<td></td>
</tr>
<tr>
<td>Trabeculated septum with coarse, parallel trabeculations.</td>
<td>Fibrous continuity between mitral and semilunar valves.</td>
</tr>
<tr>
<td>Crista supraventricularis separates tricuspid and semilunar valves.</td>
<td>No septal papillary muscles.</td>
</tr>
<tr>
<td>Papillary muscle of the conus.</td>
<td></td>
</tr>
<tr>
<td>Right branch of the bundle of His.</td>
<td></td>
</tr>
</tbody>
</table>

*The above anatomic features are the usual for the atria and ventricles. Individual chambers may lack one or more of the usual features and identification of the chamber is presumptive and based on how many of the features are present. In rare cases identification may be impossible.*

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**Figure 2. Diagrammatic representation of the features of morphologic right (RV) and left ventricles (LV). SL = septal limb of the crista supraventricularis, PL = parietal limb, PMC = papillary muscle of the conus, PA = pulmonary artery. S = septal tricuspid leaflet, MS = membranous septum, Ao = Aorta, AM = anterior mitral leaflet.**
pathologists, we find it of limited value in trying to convey information to other physicians who may also be involved in the patient's care — surgeons, house officers, referring physicians, etc. Each physician requires a lengthy explanation of both the anatomy and the 'code,' a quantum of information which may not be readily absorbed in a single sitting.

Consequently, we have sought to use Van Praagh's segmental approach but to modify it so that a) existing, easily understood terms are used; b) only a few terms are abbreviated and each abbreviated term is self-explanatory; c) the terms describe the interrelationships within a given segment as well as connections with the previous segment. The terms used are:

<table>
<thead>
<tr>
<th>Situs</th>
<th>Ventricles</th>
<th>Great Arteries</th>
</tr>
</thead>
<tbody>
<tr>
<td>solitus</td>
<td>d- or l-loop</td>
<td>d- or l-normal</td>
</tr>
<tr>
<td>inversus</td>
<td>d- or l-single RV</td>
<td>d- or l- or a-trans</td>
</tr>
<tr>
<td>asplenia</td>
<td>d- or l-single LV</td>
<td>d- or l-DORV</td>
</tr>
<tr>
<td>polysplenia</td>
<td></td>
<td>d- or l-DOLV</td>
</tr>
<tr>
<td></td>
<td></td>
<td>d- or l-malposition truncus</td>
</tr>
</tbody>
</table>

The segmental set is written as follows: situs/ventricles/great arteries, e.g., inversus/d-loop/d-trans.

The situs portion of the segmental set implies the atrial anatomy since there is viscoatrial concordance even in cases of asplenia or polysplenia.

The ventricular segment includes three terms which describe three distinct aspects of ventricular anatomy; i.e., ventricular interrelationships, the connections to the atria and the position of the ventricular portion of the heart within the thorax. As in the Van Praagh nomenclature, a "d" symbol indicates that the morphologic right ventricle lies to the right of the morphologic left ventricle, while an "l" symbol indicates that the morphologic right ventricle lies to the left of the morphologic left ventricle.

The second term in the ventricular designation has been modified from the original Van Praagh nomenclature in order to permit description of the atrial connections. If there are two ventricles and atrial-ventricular connections are of the simple type, i.e., right-sided atrium to right-sided ventricle, and left-sided atrium to left-sided ventricle, then no modification is necessary and the terms d-loop or l-loop are used. These designations are also used for cases in which one of the atrioventricular valves straddles the ventricular septum. If, however, both atria connect to one ventricle this is designated d or l-single LV or RV. The third portion of the ventricular segment describes the position of the ventricular portion of the heart within the thorax. The latter may vary considerably for a given bulboventricular loop in a given situs, e.g., a d-loop in situs solitus may have the ventricular portion of the heart in the right hemithorax (R), left hemithorax (L), or in midline (M) (fig. 3). The variable position within the thorax is the result of varying degrees of pivoting of the bulboventricular loop toward the opposite hemithorax in early fetal development. Normally, a d-bulboventricular loop pivots into the left hemithorax. Failure to complete this pivoting is particularly common with discordant loops. Clearly, the positional term may be deleted if the ventricular position is appropriate for the type of bulboventricular loop, e.g., left hemithorax for a d-loop and right hemithorax for an l-loop.

The great artery attachments to the ventricles may be described as normal (pulmonary artery from RV, aorta from LV), transposition (aorta from RV, pulmonary artery from LV), DORV (double outlet right ventricle), DOLV (double outlet left ventricle), truncus (truncus arteriosus) or malposition[21, 22] (a nonspecific term indicating an abnormal spatial relation between the aortic and pulmonary valves). Each of these is designated d- or l- to indicate the lateral in-

Figure 3. Diagram of cardiac positions with concordant and discordant bulboventricular loops. All six diagrams are of situs solitus. A, B and C each have concordant loops (d-loops); however, in A the heart is in the right hemithorax because of failure of the bulboventricular loop to pivot into the opposite hemithorax. B) Midline heart associated with partial pivoting. C) Complete pivoting into the left hemithorax, i.e., normal cardiac position for d-loop in situs solitus. D, E and F each contain discordant loops (l-loops) with complete, partial and no pivoting, respectively. Discordant loops with partial pivoting and a sagittally oriented ventricular septum tend to show mesocardia. In contrast, discordant loops with partial pivoting and a sagittally oriented septum are often more prominent on the side of the morphologic right ventricle.
terrelationships of the great arteries. In cases of transposition of the great arteries in which the aorta lies directly in front of the pulmonary artery and there is no clear d- or l-relationship, the designation a-trans (for anterotransposition) is used. The great artery lateral interrelationships need not correspond to the bulboventricular anatomy, e.g., a d-loop may be associated with great arteries in the l-position.

The following are examples of the use of this nomenclature in complex cardiac lesions.

1) **Taussig Bing**: solitus/d-loop/d-DORV with subpulmonic VSD.
2) **Corrected transposition in situs inversus**: inversus/d-loop/d-trans.
3) **Complete transposition in situs solitus with l-positioned great arteries** (an exception to the loop rule): solitus/d-loop/l-trans.
4) **Complicated double outlet right ventricle** (situs solitus with ventricular inversion and double outlet right ventricle): In this example, the d-prefix indicates that the aorta arises to the right of the pulmonary artery: solitus/l-loop/d-DORV.

Obviously, in each of these cases the associated malformations (VSD, ASD, stenosis, etc.) must be described. If the conus anatomy is the usual for a given great artery attachment, no special mention is made.

The great majority of patients do not have complex cardiac anomalies or cardiac malposition. Consequently, segmental designation is unnecessary in patients with a combination of situs solitus, a d-loop and levocardia. If, however, any aspect of cardiac malposition is present, or there is abnormal connection of the ventricles to the atria or great arteries, the entire designation is used.

Clearly, no nomenclature can be all encompassing. Rare and bizarre cases such as the criss-cross hearts described by Anderson et al. require special descriptions. Although these authors favored the terms discordant and concordant ventricles as proposed by Kirklin and coworkers, such terms are not applicable in the splenic syndromes since the symmetrical atriocoronal concordance.

We favor our method of describing cardiac anatomy because it uses only a few self-explanatory abbreviations and is quite descriptive. House officers and others not well versed in complex cardiac anatomy can understand it after only a brief explanation.

**Pathologic Material**

Sixty-five necropsy specimens exhibiting cardiac malposition were found among approximately 3,000 specimens in the Cardiovascular Registry of the United Hospitals, Miller Division, St. Paul, Minnesota. A summary of the body situses and the cardiac positions may be found in table 2. The anatomic features of the 65 cases with malposition are listed in table 3. The tabulation includes the situs and the structure of the cardiac chambers and great arteries as well as the associated intracardiac and vascular anomalies. It is apparent from table 3 that each situs is associated with a relatively small number of structural complexes but that the associated intracardiac and vascular anomalies vary markedly.

In most cases the lung fissure patterns were those expected for the situs. There were, however, sufficient variations due to incomplete and/or accessory fissures to make this an unreliable method of assessing lung morphology. In contrast the pulmonary arterial — tracheobronchial interrelationships almost always reflected the situs. This interrelationship and bronchial branching patterns were also used in determining the number of lobes.

**Situs Solitus**

There were twelve cases of dextrocardia in situs solitus. Six of these cases exhibited the normal cardiac complex of situs solitus, i.e., noninversion of the ventricles and normally connected great arteries (solitus/d-loop/d-normal). Dextrocardia in five of these cases was the result of displacement of the heart (dextroposition) associated with a hypoplastic or absent right lung. The sixth case exhibited dextrocardia (solitus/d-loop (R)/d-normal). Associated cardiac anomalies were found in four of these six cases.

The remaining six cases exhibited ventricular inversion and transposition of the great arteries (solitus/l-loop (R)/l-trans) or corrected transposition in situs solitus. In addition to the six cases of corrected transposition in situs solitus with dextrocardia, there were nineteen cases of corrected transposition with levocardia, i.e., without malposition, a distribution similar to that in previous reports. Ventricular inversion is a feature common to all cases of corrected transposition in situs solitus; however, the presence of dextrocardia or levocardia appears related to the degree of pivoting of the bulboventricular loop. The latter may best be characterized by the orientation of the ventricular septum. When dextrocardia was present, the ventricular chambers and septum were oriented in typical inverted fashion as illustrated in figure 4A-1. The ventricular septum lay oriented toward the right in a plane midway between coronal and sagittal, and the aorta was medially situated. With levocardia, the ventricles were incompletely pivoted and oriented as in figure 4A-2 with the septum perpendicular or nearly perpendicular to the coronal plane. The aorta was displaced toward the left and formed the shoulder seen in chest roentgenograms. The greater portion of the heart lay in the left hemithorax and the apparent cardiac "apex" was formed by the lateral convexity of the morphologic right ventricle. The above represent the two most common orientations of the ventricles and ventricular septum, and were the only types in this series; however, other forms have been described (see fig. 3).

**Situs Inversus**

There were thirteen cases of situs inversus which included eight with concordant loops. Of these, three had normal hearts (inversus/l-loop/l-normal), three had double outlet

| Table 2. Cardiac Position and Situs in Approximately 3000 Specimens |
|--------------------|----------------|----------------|----------------|----------------|
| Situs              | Dextrocardia   | Mesocardia     | Levocardia     | Malposition    |
| Solitus            | 12             | ~3000          | 12             |
| Inversus           | 11             | 2              | 13             |
| Asplenia           | 11             | 5              | 23             |
| Polyasplenia       | 9              | 8              | 17             |
| Total              | 65             |                |                |
right ventricle (inversus/l-loop/L-DORV), and two had
transposition of the great arteries in situs inversus
(inversus/l-loop/l-trans). There were five cases of situs
inversus with discordant loops. Three of these had
corrected transposition of situs inversus (inversus/d-loop/d-trans) and
two had double outlet of the noninverted right ventricle in
situs inversus (inversus/d-loop/d-DORV). Those cases with
discordant loops all exhibited dextrocardia, i.e., cardiac
position appropriate for that situs. The cases with discordant
loops demonstrated variation in cardiac position. The
combination of situs inversus and a d-loop may be regarded as a
mirror image of situs solitus with an l-loop, and similar but
inverted anatomic features were found.

When the discordant ventricles were associated with com-
plete pivoting, the cardiac apex was in the hemithorax op-
posite to that appropriate for the situs. In these cases the
ventricular septum was oriented toward the apex (fig. 4A-1
and B-1). With incomplete pivoting, the ventricles were
separated by a septum oriented in the sagittal plane, the
parent apex was formed by the convexity of the morphologic
right ventricle, and the characteristic shoulder was formed
by the aorta and infundibulum (fig. 4A-2 and B-2). Note
that in situs inversus the shoulder is on the right side as one might
expect.

**Asplenia**

There were 23 cases of asplenia in the present series; 11
dextrocardia, seven with mesocardia, and five with levo-
cardia. All were regarded as cardiac malpositions as no ap-
propriate cardiac position could be assigned in a
symmetrical situs.

There were 21 specimens in which lungs were attached
and each showed bilateral epipheral bronchi and bilateral right
<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Sex</th>
<th>Segmental set</th>
<th>Cardiac position</th>
<th>Cardiovascular anomalies</th>
<th>No. lobes* per lung</th>
<th>Miscellaneous</th>
</tr>
</thead>
<tbody>
<tr>
<td>45</td>
<td>2m</td>
<td>F</td>
<td>asplenia / d-loop (R) / d-normal</td>
<td>M</td>
<td>R L Total AV canal, PS,</td>
<td>3 3</td>
<td></td>
</tr>
<tr>
<td>46</td>
<td>4w</td>
<td>M</td>
<td>asplenia / d-loop (R) / d-normal</td>
<td>M</td>
<td>R L Total AV canal, PDA,</td>
<td>3 3</td>
<td></td>
</tr>
<tr>
<td>47</td>
<td>33d</td>
<td>F</td>
<td>asplenia / d-loop (R) / d-normal</td>
<td>L</td>
<td>R L Total AV canal, PDA,</td>
<td>3 3</td>
<td></td>
</tr>
<tr>
<td>48</td>
<td>3w</td>
<td>M</td>
<td>asplenia / d-loop (R) / d-normal</td>
<td>M</td>
<td>R L Total AV canal, PDA,</td>
<td>3 3</td>
<td></td>
</tr>
</tbody>
</table>

**Polysplenia**

- **R** Partial ASD, PDA, PS, primum ASD, sub AS, subvalvar AS, interrupted IVC
- **L** Partial AV canal, ASD, PDA, subvalvar AS, total AV canal

*Lung morphology as determined not only by fissure pattern but by bronchial branching patterns and relationships with pulmonary arteries.**  
**Cardiac displacement due to extrinsic forces.**

†RUPV to innominate vein; remaining pulmonary veins connect to portal vein.

†From necropsy report. Lungs and tracheobronchial tree not available for inspection.

Abbreviations: VSD = ventricular septal defect; ASD = atrial septal defect; PDA = patent ductus arteriosus; PS = pulmonic stenosis; AS = aortic stenosis; Pulm = pulmonary; AV canal = atrioventricular canal; IVC = inferior vena cava; SVC = superior vena cava; RP = right pulmonary artery; LPA = left pulmonary artery; R = right; L = left; R L or L R = right or left superior vena cava connecting to coronary sinus. If no circle the superior vena cava connects to the ipsilateral atrium.

brachial branching patterns. Fissures were a less reliable indicator of lung symmetry since not all lungs externally showed a bilateral right pattern.

The cardiac anomalies associated with asplenia were: a) anomalous systemic venous connection; b) large atrial septal defect; c) atroventricular canal; d) common ventricle or large ventricular septal defect as part of the A-V canal; e) transposition of the great arteries; f) severe pulmonic stenosis or atresia; g) anomalous pulmonary venous connection; and h) cardiac malposition.

In all cases, both atria bore resemblance to right atria (right atrial isomerism). Two large interatrial communications were commonly found. The first was an atrial septal defect of the foramen ovale type. The fossa ovalis (a right atrial structure) was usually present; however, the valve of the foramen ovale (a left atrial structure) was absent. The second interatrial communication was the atrial portion of the complete atroventricular canal. Both defects were usually quite large with only a strand of septal tissue separating them so that functionally there was a common atrium (fig. 5).

There were two ventricles in 14 cases and a single ventricle in nine cases. All cases had a complete atroventricular (A-V) canal. When two ventricles were present, the intraventricular portion of the A-V canal was sufficiently large to result in a hemodynamic common ventricle with a rudimentary septum. When two ventricles were present, there was commonly inversion of the ventricles (nine of fourteen cases).

Severe pulmonic stenosis or atresia was found in all but two cases and was usually of both the subvalvar and valvar types. Transposition of the great arteries was found in 19 cases, double outlet right ventricle in three cases, and normally related great arteries in only one case.

Anomalous systemic venous connection was also a common feature. Twenty cases exhibited bilateral superior vena cavae and in each case right and left superior vena cavae connected directly to their respective atria. The inferior vena cava terminated in either atrium.

Anomalies of pulmonary venous connection occurred in 16 of the 23 cases of asplenia. The majority of these were total anomalous pulmonary venous connection. The sites of connection of the anomalous pulmonary veins were the portal system or a superior vena cava, i.e., anastomoses with vessels derived from the umbilicotvillarine anterior cardinal systems, respectively.

Thirteen of the cases of asplenia in the present series (cases 28–33, 35, 37, 40, 43–45 and 48) have been reported previously.*

**Polysplenia**

There were 17 cases of polysplenia in the present series; nine with dextrocardia and eight with levocardia.

Some, if not all, of the features of bilateral left-sidedness were found in each case. These included a tendency toward bilateral left atria and bilateral left lungs. There were 14 specimens in which the lungs were available and 12 showed bilateral hyparterial bronchi and bilateral left bronchial branching patterns. The remaining two specimens showed bilateral eparterial bronchi and bilateral right bronchial
branching patterns. Lung fissures varied somewhat but in most cases were of the bilateral left pattern. As in the asplenia group, lung fissures were less reliable than pulmonary artery-bronchial relationships in evaluating lung symmetry. The abdominal organs displayed less symmetry than in asplenia; malrotation of the bowel was present in at least three cases.

The cardiovascular manifestations of polysplenia included: a) anomalous systemic venous connection; b) anomalous pulmonary venous connection; c) atrial septal defects; d) ventricular septal defects; e) double outlet right ventricle; f) left-sided obstructive lesions; and g) cardiac malposition. In contrast to asplenia, transposition of the great arteries and pulmonic stenosis were unusual.

Anomalies of the systemic venous drainage may involve the superior and/or inferior vena cavae. In eight cases of polysplenia, bilateral superior vena cavae connected directly to their respective atria, while in two cases the second superior cava connected to the coronary sinus. Nine cases also exhibited interruption of the hepatic portion of the inferior vena cava with drainage into a superior vena cava by the hemiazygos or azygos system. When interruption of the inferior vena cava was present, the hepatic veins drained directly into one or both atria. The atrium into which the inferior vena cava drained, either directly or by the azygos system, received most of the systemic venous return and we have designated this the systemic venous atrium.

Often both atria morphologically resembled left atria (left atrial isomerism) and each contained a smooth-walled atrial appendage. Both sides of the atrial septum frequently bore resemblance to the normal left atrial aspect of the atrial septum (septum primum) and in these cases the foramen ovale was absent (fig. 6).

Anomalous pulmonary venous connection was found in 15 cases. Eleven were partial with the right pulmonary veins connecting directly to the right-sided atrium. There were four cases of total anomalous pulmonary venous connection and in each the veins connected with the systemic venous atrium directly or through a confluen.

Defects of the atrial and/or ventricular septum were present in all 17 cases of polysplenia. There were 12 specimens with ventricular communications; in five the ventricular septal defect was part of an atrioventricular canal and in seven it was a discrete lesion. Fifteen specimens had one or more defects of the atrial septum. In one the defect was of the os-tium primum type, and in two the atrial septum was completely lacking. Twelve specimens exhibited atrial septal defects which were the result of fenestrations or deficiencies
in the valve of the foramen ovale and/or absence of both limbi of the fossa ovalis. Seven cases also had patency of the ductus arteriosus. The above communications, as well as the anomalous pulmonary venous connections, are lesions which, in the absence of pulmonary stenosis, would result in pulmonary overcirculation.

Pulmonic stenosis was uncommon, being found in only two cases. In contrast, left-sided obstructive lesions were common. There were five cases with aortic outflow obstruction and two with aortic coarctation.

Twelve of the cases of polysplenia in the present series (cases 49-52, 56, 57 and 60-65) have been reported previously.11

Discussion

The anatomic findings in the present series were very similar to those of Lev and associates96 and Van Praagh and associates,44 although neither study classified polysplenia as a distinct situs. Tetralogy of Fallot with cardiac malposition was virtually absent in both these series as well as in our cases. This differed considerably from the earlier work of Arcilla and Gasul44 which reported seven cases of tetralogy; however, only two of their seven cases of tetralogy were examined at necropsy. It is possible that some of the cases of tetralogy may have had ventricular inversion (i.e., "corrected transposition" with pulmonic stenosis and ventricular septal defect).

The large proportion of cases of malposition associated with splenic anomalies is noteworthy and is in agreement with the findings of others.19, 80, 81 It is quite likely that many of the cases previously reported as "mixed situs" or "incomplete situs inversus" were, in reality, examples of splenic anomaly syndromes, particularly polysplenia.

The findings in the asplenia and polysplenia cases are similar to those reported by Van Mierop12 and Rose and co-workers;43 however, the latter reported an unusually high incidence of pulmonary outflow obstruction in polysplenia (four of 12 cases). Pulmonic stenosis was present in only two of 17 cases in the present series. In contrast, seven of 17 cases in the present series showed aortic outflow obstruction or coarctation.

Ivemark reported four cases of type IV truncus arteriosus in association with asplenia;83 truncus was not present in any of the asplenia cases in this series as well as others.14, 90, 91

The high incidence of cardiac anomalies in cases of situs inversus (11 of 13 cases) exceeded that found by Arcilla and Gasul44 and contrasted sharply with the findings of Keith and co-workers.49 Grant,90 and Torgersen.96 The latter study was based, in part, on screening chest roentgenograms of persons over 15 years of age and excluded cases with severe anomalies which caused an early death. The present necropsy study is probably weighted to the opposite extreme. The true incidence of cardiac anomalies in situs inversus probably lies somewhere in between.

Similarly, the incidence and severity of congenital cardiac anomalies associated with polysplenia is probably greater in necropsy series than in catheterization series. Although there were no cases of polysplenia without cardiac anomalies in the present series, the authors have catherized two patients with probable polysplenia, dextrocardia, an interrupted inferior vena cava and no intracardiac anomalies. Individuals with polysplenia, levocardia and no intracardiac anomalies might not even come to a physician's attention.

Clinical Correlates of Anatomic Features

Identifying the Situs

The situs of the individual patient is best determined by chest and abdomen roentgenograms with additional laboratory studies (table 4).

In situs solitus, the viscera are normally situated while in situs inversus inverted viscera are evident. In asplenia, the abdominal viscera are very symmetrical with a horizontal liver and a stomach bubble which tends to lie toward the midline.98 In polysplenia, the tendency toward symmetry is also present but is not as striking as in asplenia. As both lungs are often bilobed in polysplenia, the presence of a minor lobe fissure on chest roentgenograms is strong evidence against polysplenia. In contrast, the finding of the upper abdominal portion of the aorta on the side opposite the stomach is strong evidence for polysplenia.97 Roentgenograms showing the symmetrical bronchial patterns in the splenic anomaly syndromes have also been helpful.89 Malrotations of the bowel are common in both asplenia and polysplenia.90 Radioisotopic scanning may demonstrate absent or multiple spleens.95, 40, 41
As the atria almost always follow the situs, the direction of the P vector on the electrocardiogram may be of some value in determining the situs (fig. 7). In situs solitus, atrial depolarization proceeds from the sinoatrial node toward the left and inferiorly resulting in an upright P in I, II, III, aV₃, and aV₅. In situs inversus, the P vector is from left to right and inferiorly with an upright P in II, III, aV₉, and inverted P in I and aV₅. In asplenia, both atria are morphologic right atria and each may have a sinoatrial node. Consequently, either of the above P vectors may be present and both may be present in the same patient at different times. In polysplenia, ectopic low atrial pacemakers are common with negative P waves in II, III and aV₉. This may be the result of bilateral left atria and absent sinoatrial nodes. We know of no histologic studies of sinoatrial nodes in polysplenia.

Situs Solitus

Cardiac malpositions in situs solitus in this series were the result of either cardiac displacement (dextroposition), dextrotransposition, or a discordant loop (solitus/l-loop).

Dextroposition may be recognized readily by an asymmetric thorax and decreased breath sounds on the right as well as an absent or hypoplastic right lung in chest roentgenograms.

The physical findings in corrected transposition in situs solitus (solitus/l-loop/l-trans) are the result of the abnormally positioned semilunar valves and the associated cardiac anomalies. The anterior and leftward position of the aortic valve results in a very loud aortic closure on auscultation which is usually maximal at the upper left sternal edge. The second sound is usually single, but occasionally a split is heard in the vicinity of the displaced pulmonary valve, i.e., the mid or right sternal edge. When pulmonic stenosis is present, the murmur may be maximal retrosternally or to the right of the upper sternum. The murmur of the left atrioventricular valve insufficiency may be present in patients with Ebstein's malformation of this valve.

Asplenia

The numbers in parentheses are approximate incidences based on information in the literature as well as the authors' clinical experience. (R = right; L = left; SVC = superior vena cava; IVC = inferior vena cava; APVC = anomalous pulmonary venous connection; CHF = congestive heart failure.)

![Table 4. Clinical and Laboratory Features of the Four Body Configurations](image-url)

**TABLE 4. Clinical and Laboratory Features of the Four Body Configurations**

<table>
<thead>
<tr>
<th>Presenting symptoms</th>
<th>Solitus</th>
<th>Inversus</th>
<th>Asplenia</th>
<th>Polysplenia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cyanosis (~95%)</td>
<td>CHF (~5%)</td>
<td></td>
<td>Cyanosis (~20%)</td>
<td>CHF (~80%)</td>
</tr>
<tr>
<td>Roentgenograms – Abdomen</td>
<td>Normal</td>
<td>Inverted</td>
<td>Horizontal liver</td>
<td>Malrotation</td>
</tr>
<tr>
<td>Thorax</td>
<td></td>
<td></td>
<td>Midline stomach</td>
<td></td>
</tr>
</tbody>
</table>
| Splenia, ectopic anomalies. The anterior normally positioned semilunar valves and the associated cardiac anomalies. The anterior and leftward position of the aortic valve results in a very loud aortic closure on auscultation which is usually maximal at the upper left sternal edge. The second sound is usually single, but occasionally a split is heard in the vicinity of the displaced pulmonary valve, i.e., the mid or right sternal edge. When pulmonic stenosis is present, the murmur may be maximal retrosternally or to the right of the upper sternum. The murmur of the left atrioventricular valve insufficiency may be present in patients with Ebstein's malformation of this valve.

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The roentgenographic features of corrected transposition in situs solitus (solitus/d-loop/d-trans) are quite variable. The two general forms of cardiac silhouette have been described earlier. The pulmonary vascularity is related to the degree of pulmonic stenosis and/or shunting at the ventricular level. Even in the absence of pulmonic stenosis, the main pulmonary artery may not be evident because of its medial position.

The bundle branches also follow the respective ventricles and this probably explains the electrocardiographic features of corrected transposition. In d-loops (including cardiac displacement) septal depolarization proceeds from left to right and a Q wave is recorded in the left precordial leads. With l-loops, septal depolarization proceeds from right to left and a Q wave is frequently recorded in the right but not over the left precordium. As the degree of ventricular rotation and, therefore, septal position may vary, the Q waves may not be evident in V6 but may be present in the more rightward chest leads. This feature is not constant and similar Q wave patterns may be found with right ventricular hypertrophy without ventricular inversion. In addition, atrioventricular conduction defects are common in corrected transposition.

The anatomic positions of the valves of the great arteries result in unusual catheter positions during cardiac catheterization. The location of the pulmonary valve orifice behind the venous atrioventricular valve and subpulmonic stenosis may preclude entering the pulmonary artery. Even when the main pulmonary artery is entered the acute angulation of the catheter frequently prevents advancement to the pulmonary arterial wedge position unless a balloon tipped catheter is used.

**Situs Inversus**

Cases of situs inversus with concordant ventricles show dextrocardia as a constant feature and this is evident by physical and roentgenographic examination as well as “mirror image” progression of precordial QRS complexes on the electrocardiogram. There is a rightward inferior P axis (fig. 7) and no findings suggestive of cardiac disease.

The physical findings in corrected transposition in situs inversus (d-loop/d-trans) include a very loud aortic closure maximal at the upper right sternal edge and the features of associated anomalies. The two general forms of cardiac silhouette seen on roentgenograms have been described above (fig. 4). When a “shoulder” is present in corrected transposition in situs inversus it is on the right heart border, i.e., the same side as the apparent cardiac apex. When the heart is left-sided, a “shoulder” is notably absent. The electrocardiographic features are the rightward P axis of situs inversus and septal depolarization proceeding from left to right.

**Asplenia**

The clinical picture begins with cyanosis in the neonatal period. Survival beyond infancy is unusual and serious infections such as meningitis are not rare. With the exception of a horizontal liver, the physical findings are not peculiar to asplenia.

The roentgenographic features are those of pulmonary undercirculation, a markedly symmetrical liver, a midline stomach bubble, malrotation of the bowel, symmetry of the tracheobronchial tree, and decreased pulmonary vascularity. Anomalies of the liver, spleen, and associated cardiac malformations are common.

As two sinoatrial nodes may be present, the electrocardiogram may show either an inferior rightward or inferior leftward P axis. The same patient may show both at different times. Although the lungs may be affected beyond the liver, spleen, and pancreas, the physical findings are not suggestive of asplenia.

**Polysplenia**

The clinical features of polysplenia are in marked contrast to those of asplenia. Cyanosis is usually absent or minimal and congestive heart failure is common in those patients with polysplenia. Hepatic and cardiac anomalies are not distinctive for polysplenia. Chest and abdomen roentgenograms show a variety of hepatic and cardiac positions; however, mesocardia is rare. Focusing attention on the position of the heart and lungs may result in cases of polysplenia being missed or categorizing them as situs inversus or partial situs inversus. Additional roentgenographic findings that favor the diagnosis of polysplenia are bilateral left bronchial pattern, absence of a minor lobe fissure in both lungs and malrotation of the bowel. A feature peculiar to polysplenia is the upper abdominal aorta lying on the side opposite the stomach bubble. Low atrial pacemakers are found frequently on electrocardiogram. Peripheral blood smears show no Heinz or Howell-Jolly Bodies (unpublished observations). Although more than 100 cases of polysplenia have been documented there are no reports of unusual susceptibility to infection. Recent publications added six cases of extraperitoneal biliary atresia to the seven previously reported cases.

At cardiac catheterization interruption of the inferior vena cava with azigos continuation is strong evidence for polysplenia and should alert the cardiologist to this possibility even in the absence of cardiac or hepatic malposition. Symmetry of the pulmonary arteries may be apparent in the posterior-anterior projection but the “bilateral-left” configuration is best verified in the lateral projection. In this view both lower lobe pulmonary arteries lie in the same coronal plane and are posterior to the bronchi.

Although there are several clinical indicators for polysplenia, none are pathognomonic. Unless a radioisotope splenic scan shows an irregular splenic mass suggestive of polysplenia the diagnosis is usually presumptive and its likelihood depends on how many of the above.
features are present. A definitive diagnosis occasionally is made at laparotomy for bowel obstruction or biliary atresia.

**Embryologic Considerations**

**Situs**

The anatomic features of the viscera in each of the situses may be explained readily if one hypothesizes that there are separate factors controlling the development of morphologic right and morphologic left structures from paired lateral isomers. When both factors are present, the resulting body configuration is either situs solitus or inversus, depending on the factors’ interrelationships. When the factors controlling right morphology are present bilaterally, asplenia syndrome may be expected to occur and with duplicate left factors, polysplenia syndrome. Although many organs show symmetry in asplenia and polysplenia, others do not. The lungs, tracheobronchial tree, atria and liver are symmetric or tend toward symmetry while the ventricles and great arteries are asymmetric. The symmetric organs develop from midline anlage which have undergone a sagittal division into right and left isomers. Whatever factors control right-left morphology probably influence these isomers to be symmetric in each of the splenic syndromes. On the other hand, structures which result from cephalad-caudal or coronal division are not symmetric. The great arteries are the result of a coronal division of the truncus arteriosus into anterior and posterior portions (aorta and pulmonary artery, respectively). Similarly, the right and left ventricles develop from anlage that are at least partially cephalad-caudal in the cardiac tube. In each of these instances, the paired structures are not derived from lateral isomers and might not be expected to be influenced by factors controlling right-left morphology.

The gastrointestinal tract is asymmetric but has a specific orderly arrangement in both situs solitus and situs inversus. This orderly arrangement within the body might require the influence of both right and left factors. The presence of bilateral right or left factors might result in a disorderly arrangement of the bowel, i.e., malrotation.

The splenic anlage makes its appearance relatively late in gestation (i.e., 30 days). Normally, the splenic anlage appears as a small bud on the left side of the dorsal mesogastrium. Van Mierop has examined the splenic tissue of patients with polysplenia at necropsy and found spleens on each side of the dorsal mesogastrium. The latter lends support to the concept of bilateral left-sidedness.

Conversely, absence of the spleen in asplenia syndrome is probably another manifestation of bilateral right-sidedness with either suppression or agenesis of the splenic anlage as part of the developmental complex. On the other hand, isolated absence of the spleen, i.e., without visceral abnormalities, is probably the result of agenesis of the splenic anlage as an independent event. There is one such case in the Cardiovascular Registry of the United Hospitals, Miller Division.

The significance of accessory spleen in these developmental complexes is unclear. Accessory spleens are a fairly common finding at routine postmortem examinations and usually are not associated with other congenital visceral or cardiac anomalies. Case 59, in the present series, is an exception. Lung fissures, atrial morphology and atrial septal morphology were similar to that in polysplenia; however, tracheobronchial and pulmonary arterial relationships were typical of bilateral right-sidedness.

**Cardiac Position**

The d- or l- bulboventricular loop determines the positions of the ventricles relative to each other and to the atria. The position of the heart within the thorax is dependent not only on the type of bulboventricular loop but also on the degree of pivoting of the ventricular portion of the heart. Normally, at four weeks when a d-loop forms the ventricles, the ventricular portion of the heart is still directed toward the right with the ventricles situated side by side. At nine weeks, the ventricular portion of the heart pivots toward the left, placing the heart in the left hemithorax and the right ventricle anterior to the left ventricle. Conversely, with an l-loop the ventricular portion pivots toward the right hemithorax. In our experience, when the loop was concordant with the situs, the pivoting was complete in almost all cases. The only exception was the case of solitus/d-loop (R)/d-normal. With discordant loops, the pivoting is frequently incomplete resulting in the varying cardiac positions found in corrected transposition of situs solitus (solitus/l-loop/l-trans), as well as corrected transposition of situs inversus (inversus/d-loop/d-trans). This same principle can be applied to cases of asplenia and polysplenia. The symmetry of these situses precludes designating discordant loops. Therefore, it is not surprising that each type of bulboventricular loop in each of these situses showed variation in degrees of pivoting and cardiac position (table 5).

**Pulmonary Veins**

The differing patterns of pulmonary venous connection found in asplenia and polysplenia syndromes have been related to the type of atrial isomerism found in each.

### Table 5. Cardiac Position Related to Situs and Bulboventricular Loop

<table>
<thead>
<tr>
<th>Situs</th>
<th>Loop</th>
<th>Total no. cases</th>
<th>Dextrocardia</th>
<th>Mesocardia</th>
<th>Levocardia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Solitus</td>
<td>d-loop</td>
<td>~3000</td>
<td>6†</td>
<td>(3000)†‡</td>
<td></td>
</tr>
<tr>
<td></td>
<td>l-loop*</td>
<td>15</td>
<td>6</td>
<td>(19)†‡</td>
<td></td>
</tr>
<tr>
<td>Inversus</td>
<td>d-loop*</td>
<td>5</td>
<td>3</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>l-loop</td>
<td>8</td>
<td>4</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Asplenia</td>
<td>d-loop*</td>
<td>8</td>
<td>4</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td></td>
<td>l-loop*</td>
<td>7</td>
<td>7</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Polysplenia</td>
<td>d-loop*</td>
<td>12</td>
<td>5</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td></td>
<td>l-loop*</td>
<td>5</td>
<td>4</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

*Discordant bulboventricular loops.
†5 cases of cardiac displacement and 1 of incomplete pivoting of the ventricles, i.e., solitus/ d-loop (R).
‡( ) indicates specimens without cardiac malposition and not included in this study.
asplenia there are bilateral right atria and consequently bilateral absence of the common pulmonary vein.\textsuperscript{6} As a result, the sites of connection of the anomalous pulmonary veins in asplenia are the superior vena cavae or the portal system (fig. 8). As there are two left atria in polysplenia, a common pulmonary vein may develop from either or both\textsuperscript{19} and the pulmonary veins may connect directly to either or both atria. Partial anomalous pulmonary venous connection may be the result of a common pulmonary vein arising from each of the morphologic “left” atria, whereas total anomalous pulmonary venous connection to the venous atrium may be the result of a common pulmonary vein developing from that atrium (fig. 9).

In cases of polysplenia where the pulmonary veins connect directly to both atria, they do so in a distinctive fashion. The right veins connect to the right-sided atrium and the left veins to the left-sided atrium. Van Mierop has pointed out that the sites of connection are not at the intercaval portions derived from the sinus horns but rather more medially and adjacent to each side of the atrial septum.\textsuperscript{19} The latter connection suggested that the partial anomalous pulmonary venous connection in polysplenia was the result of bilateral common pulmonary veins in bilateral left atria.

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