Computed Tomography Pulmonary Angiography in Evaluation of Congenital Heart Diseases

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ABSTRACT

Congenital heart disease (CHD) still accounts for significant morbidity and mortality, despite major advances in diagnosis and management. A significant proportion of CHD is still undiagnosed at birth emphasizing the need for a detailed history and meticulous clinical examination. CHD is common and early detection and comprehensive functional and physiological evaluation is very important. Usually echocardiography is the initial diagnostic modality for patients with suspected CHD; in some patients this modality can be limited in its ability to delineate great artery and intracardiac anomalies, pulmonary veins, and coronary arteries. Magnetic resonance imaging is an excellent tool due to its non-ionizing nature and exquisite anatomic and functional capabilities. Computed tomography (CT) pulmonary angiography can be used to assess complex cardiovascular anatomic features both before and after surgery and of a variety of post-treatment complications. In CHD it is very important to have extensive knowledge of cardiovascular anatomy, physiology, and surgical techniques. Recent developments in CT technology primarily by reducing the cardiac motion and the radiation dose usage in CHD evaluation have helped expand the indications for CT usage. It has become a very useful complementary tool to guide the proper medical and surgical management. The purpose of the article is to review the role of CT pulmonary angiography in the evaluation of congenital cardiovascular disease with emphasis on its type, nature and anatomic details.

Keywords: Cardiac anomalies, congenital heart disease, computed tomography pulmonary angiography, imaging

INTRODUCTION

The human heart is a scaffold of complex and intricate relationships between its different components. Consequently, errors in its embryogenesis, present in many different combinations and their accurate diagnostic characterization requires a profound proficiency on the part of both cardiologists and radiologists. The variety of presentations of congenital heart disease is so perplexing that every now and then we encounter a case which defies several classification systems that has been designed to describe these diseases. Congenital heart diseases have been probed by all the existing imaging modalities from plain radiography, angiography, radionuclide imaging to advanced 2D imaging techniques such as echocardiography, computed tomography (CT) and magnetic resonance imaging (MRI), and every of these modalities have an important role. Thoracic radiography, often considered banal in modern day practice, may provide the first indication that heart disease is present and if read carefully provide a generalized notion of the type of these lesions rather than a specific diagnosis. Invasive angiography is limited by catheter related complications, overlapping of adjacent cardiovascular structures, inability to assess systemic and pulmonary vasculature simultaneously and relative high dose of radiation dose and iodinated contrast material and reserved mainly for invasive pressure measurement. Echocardiography provides a wealth of both anatomical and dynamic physiological information without significant patient discomfort or risk. It has, therefore, become a diagnostic ritual for the first hand analysis of patient suspected for congenital heart lesions or any heart disease for that matter. Furthermore,
echocardiography is being used in many centers intraoperatively to assess immediate post repair global pump function and provide immediate evaluation of results. However, transthoracic echocardiography is limited by certain blind spots such as ascending aorta and the arch of the aorta, the distal pulmonary arteries, the right ventricle, and the pulmonary veins. Still, in modern day practice, echocardiography is most well-suited for initial assessment, and it would not be an overstatement to say that most of the evaluation start with echo and end with echo. If it needs to go further, the choice of investigation lies between CT and MRI. Although MRI is extremely meticulous in detailing the anatomical and functional character of the heart, it is time-consuming and coerces the need to sedate and intubate the patients especially in pediatric age group. CT, on the other hand, is extremely fast and has widespread availability but exposes patients to significant radiation burden. A trade-off between advantages and disadvantages between these two modalities must be made, and a diagnostic protocol must be individualized from patient to patient basis. Modern CT work stations are equipped with multiplanar reformation techniques which is necessary for accurate assessment of various vessels because vessels through their course change their axis and can be optimally evaluated in a different plane. The analysis of congenital heart lesions is best achieved by systematic segmental approach which divides heart into three main segments, i.e., atria, ventricles and the great vessels and the connections between these chambers, i.e., atrio-ventricular and ventriculo-atrial connections. Careful step by step evaluation of viscera-atrial situs, cardiac chambers, their connections, and associated malformations should be done to complete the CT study. This article describes the various CT features of various congenital heart diseases which will help the radiologists to interpret these complex anomalies.

**EXTRACARDIAC ANOMALY**

**Interrupted aortic arch**

In this defect, a portion of the aortic arch is absent. There are three types of the aortic arch interruption.1

1. **Type A**: The interruption occurs just beyond the left subclavian artery
2. **Type B**: The interruption occurs between the left carotid and the left subclavian artery. It is the most common type of interruption
3. **Type B2**: The right subclavian artery originates as fourth anomalous branch of the aortic arch along with the Type B interruption
4. **Type C**: The interruption occurs between the innominate and the left carotid artery.

The lesion mostly is associated with patent ductus arteriosus (PDA) and ventricular septal defect (VSD). Association with Di-George syndrome (with right-sided descending aorta) and double outlet right ventricle (DORV) with VSD (Taussig–Bing malformation) has been also reported.2,3

CT is invaluable in surgical planning and a complete evaluation should cover4,5

- Associated cardiac anomalies
- The distance between the proximal and distal segments
- The size of a PDA
- The narrowest dimension of the left ventricular outflow tract

CT can be used to assess the presence of vascular collaterals. A patient adequately compensated by rich collaterals can survive to adulthood asymptotically without any surgical intervention. However, complications like aortic dissection sometimes develop which can be potentially life threatening.6

**Coarctation of the aorta**

First described by Morgagni in 1760, coarctation of aorta is congenital narrowing of descending aorta or distal aortic arch. It is usually caused by fibromuscular ridge extending into lumen just distal to left subclavian artery adjacent to the insertion of ligamentum arteriosus (juxtaductal coarctation) or tubular narrowing of the aortic isthmus.8,9 Coarctation is commonly associated with PDA (Figure 1) and VSD, and the triad of these three has been named as coarctation syndrome. Bicuspid aortic valve and Turner syndrome are other associations.3

![Figure 1. Computed tomography pulmonary angiography study sagittal reformatted image of a pre-ductal coarctation of the aorta showing significant luminal narrowing in descending aorta with patent ductus arteriosus and lack of collaterals.](image-url)
Multidetector CT (MDCT) with multiplanar and volume rendered reconstructional images can accurately detect the coarctation along with the evaluation of the degree of stenosis and collateral vessels (Figure 2) (collateral epigastric, intercostal, thoracoacromial, vertebral, and anterior spinal arteries). Although CT is inferior to MRI, which is exquisitely sensitive in providing hemodynamic information in cases of mild stenosis, CT is invaluable in postsurgical evaluation in which MRI imaging is often suboptimal due to magnetic susceptibility artifacts by metallic stents. CT can detect other complications of coarctation involving the ascending and transverse aorta; the subclavian, radial, brachial, and carotid arteries; and the retinal vascular bed such include collateral arteries, vascular rings, and bicuspid aortic valve, dissecting aneurysms, cerebral aneurysms, and decreased left ventricular interpapillary distance. CT has also proved to be an excellent modality for stent follow-up and detection of stent fracture and secondary aortic rupture.

Truncus arteriosus

It is a single arterial vessel that originates from the heart and supplies the systemic, pulmonary, and coronary circulation. The trunk bestrides a high VSD. An association with Di-George syndrome and chromosome 22q11 deletion has been reported. The classification of truncus arteriosus is based on the branching pattern of the pulmonary artery. The original and more popular classification system was devised by Collett and Edwards in 1949, which divides this anomaly into four types:

- Type 1: Common pulmonary artery arising from truncus
- Type 2: Right and left pulmonary artery arise separately from the posterior part of truncus
- Type 3: Separate origin of the pulmonary arteries from the lateral aspect of the truncus
- Type 4: Neither pulmonary arterial branch arising from the common trunk (pseudo-truncus), is now recognized to be a form of pulmonary atresia with VSD rather than truncus arteriosus.

In 1965, Van Praaghs modified this classification system also includes four primary types:

- Type A1: Identical to the Type I of Collett and Edwards
- Type A2: Separate origin of the branch pulmonary arteries from the left and right lateral aspects of the common trunk
- Type A3: Origin of one branch pulmonary artery (usually the right) from the common trunk, with other lung supplied either by collaterals or a pulmonary artery arising from the aortic arch
- Type A4: Coexistence of an interrupted aortic arch.

Occasionally, one lung is supplied by truncus, and the other lung is supplied by pulmonary artery arising from the right ventricle. This anomaly is called “hemitruncus” and is often associated with pulmonary stenosis. CT demonstrates the truncus arteriosus and associated abnormalities easily.

PDA and other aorto-pulmonary shunts

The fetal ductus arteriosus bridges the proximal left pulmonary artery just distal to the pulmonary artery bifurcation with the proximal descending aorta immediately distal to the origin of left subclavian artery. If the ductus arteriosus does not close spontaneously within 1st day of life, there is a significant left to right shunt from the descending aorta to the pulmonary arteries. PDA is often found in many cyanotic pulmonary obstructive lesions such as pulmonary atresia and are life sustaining in such cases. Association of PDA with mosaic ring chromosome 6 has been reported along with hypoplasia of aortic valve and aortic arch. The lesion is easily detected by echocardiography and role of CT is limited to assess the complications of PDA such as ductus arteriosus aneurysm. Accurate delineation of size and shape of ductus along with detection of track like ductus calcification can be reliably provided by CT which aids making a decision whether as to perform surgery or transcatheter ductus closure.

Other less frequent aortopulmonary shunts are aortopulmonary window, aortic sinus fistula, coronary artery arising from the pulmonary artery, coronary artery fistula into the right heart, postsurgical shunts, and arteriovenous
fistula of the lung and chest wall. Aortopulmonary window is a large connection between ascending aorta and main pulmonary artery with the presence of separate well-formed aortic and pulmonary artery valve, thus differentiating it from truncus arteriosus.

Aortic sinus fistula is usually a complication of funnel-shaped aneurysmal dilatation of one of the sinuses of valsalva. More commonly right coronary or noncoronary sinuses rupture into the right atrium or right ventricle. Differential diagnosis includes Marfan’s syndrome which causes diffuse dilatation of the sinuses.\textsuperscript{3,15}

**Total anomalous pulmonary venous connection**

Total anomalous pulmonary venous connection is a connection of all the pulmonary veins to the right atrium either directly or indirectly via the superior or the inferior vena cava (IVC) with the pulmonary veins usually forming a confluence before entering systemic venous system behind left atrium. There are mainly four patterns:\textsuperscript{3}

1. Supracardiac drainage: Connections are to the left innominate vein, superior vena cava (SVC), or azygos vein. Veins are usually prominent, even aneurysmally dilated to produce “figure of eight” or “snowman” appearance on radiography
2. Cardiac drainage: Connections are to the right atrium, either directly or via coronary sinus
3. Infracardiac drainage: Connections are to the portal vein its branches, ductus venosus, or hepatic vein. This variety is usually obstructed to high vascular resistance of liver and compression at diaphragmatic hiatus

CT is a valuable tool for follow-up evaluation after surgery and to anticipate the need for reintervention. Pulmonary venous stenosis is a frequent complication of surgery and can be easily recognized by CT.\textsuperscript{16}

**Partial anomalous pulmonary venous connections (PAPVC)**

In PAPVC, at least one pulmonary vein drains to a location other than the left atrium, most commonly into SVC or right atrium and is typically seen with atrial septal defect (ASD), often and sinus venosus defect (Figure 3). Patients are usually only mildly symptomatic due to a small amount of shunt. Scimitar syndrome (hypogenetic lung or pulmonary venolobar syndrome) is a rare form of PAPVC in which right lower and sometimes middle lobe forms a vein that runs downward and medially in crescent course passing through diaphragm to enter IVC. Other findings observed in patients with scimitar syndrome include a small ipsilateral hemithorax, cardiac dextroposition and varying degree of pulmonary artery hypoplasia or aplasia. There are often systemic collaterals to right basal lung segments from abdominal aorta and basal bronchiectasis. CT is very useful in diagnosis and characterization of these lesions.\textsuperscript{3,8,17}

**Pulmonary artery sling**

In pulmonary artery sling, an aberrant left pulmonary artery arises from the proximal right pulmonary artery, passes between the trachea and esophagus, and enters low into the left hilum, thus forming a sling around the right main bronchus and trachea. Abnormalities of the tracheobronchial tree are common association.\textsuperscript{8}

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**CARDIAC ANOMALIES**

**Tetralogy of fallot (TOF)**

TOF represents the most common cyanotic cardiac disease, accounting for almost 12% of all congenital heart anomalies. The classic tetrad of manifestations i.e., pulmonary outflow tract stenosis, VSD, aortic overriding, and right ventricle hypertrophy, are all consequence of eccentric truncus septation by conal septum resulting in pulmonary valve hypoplasia along with hypoplasia of main pulmonary artery and origin of right and left pulmonary artery. The conotruncal septum is anteriorly displaced, and its lower part fails to fuse with the interventricular septum leading to high VSD (Figure 4). Anterior septal displacement...
also causes aorta to override the VSD. VSD is large and nonrestrictive leading to equal pressure in both ventricles. Pulmonary stenosis can variably involve:

- Stenosis below the pulmonary valve, between displaced infundibular septum and anterior wall of the right ventricle, also accentuated by hypertrophied septal muscle bundles. Periods of infundibular spasms often accompany the cyanotic spells of the affected children
- Stenosis of abnormal pulmonary valve, with hypoplastic annulus and thickened, fused, and doming leaflets producing valvular stenosis (Figure 5)
- Stenosis of sinotubular junction
- Stenosis or atresia of origin of the right and left pulmonary arteries. Even in the absence of this branch stenosis, right pulmonary obtains preferential flow owing to its orientation

- Atresia of the pulmonary valve with VSD is an extreme form of TOF.

About 20% of patients (more frequent with severe pulmonary stenosis) have a right aortic arch, usually with mirror-image branching (Figure 6). Association with double aortic arch has also been reported. Pulmonary flow may be maintained by PDA or a large major aorta pulmonary collateral arteries (MAPCAs) arising from descending aorta, brachiocephalic artery or coronary arteries but never from ascending aorta (Figure 7). These large arteries may be visible on radiographs but are more clearly depicted on CT. Missing these arteries preoperatively is a potential cause of torrential intraoperative bleed. Surgical anatomy of right ventricle outflow tract and pulmonary valve is seldom reliably assessed by echocardiography and can

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**Figure 4.** Pulmonary angiography axial section showing a large ventriculo-septal defect (black arrow).

**Figure 5.** Pulmonary angiography sagittal reconstructed image showing infundibular pulmonary stenosis and hypoplastic annulus (solid black arrow) in a case of tetralogy of fallot.

**Figure 6.** Pulmonary angiography axial section showing right side aortic arch with mirror image branching (solid white arrow) in a case of tetralogy of fallot.

**Figure 7.** Computed tomography pulmonary angiography sagittal reconstructed image showing major aorto-pulmonary collaterals from descending thoracic aorta as a result of underlying congenital heart disease (solid black arrows) maintaining pulmonary flow.
be achieved noninvasively by CT along with multiplanar reconstructional images with considerable accuracy. The ratio of the diameter of the aorta (measured at diaphragmatic level) to the sum of the right and left pulmonary arteries has been used to triage ideal candidates for primary corrective surgery. Descending aorta/(right + left pulmonary artery) ratio ≤0.6 indicate pulmonary arteries of sufficient size for primary correction, while higher values are an indication for palliative surgeries.\textsuperscript{18} CT may resolve other considerations about additional VSD, coronary artery course, and MAPCA.\textsuperscript{19,20}

ASD

Imaging of ASDs by CT requires good intra-cardiac opacification of left heart for detection of left to right shunt which is achieved using a biphasic protocol consists of an initial contrast bolus at a rate of 4-7 ml/s, followed by 40-50 ml of saline, to flush out the contrast from the right heart.\textsuperscript{21} As the shunting predominantly occurs in late diastole, electrocardiography (ECG) gated CT acquisition is preferred for optimal imaging. There are four types of ASDs, secundum ASDs (most frequent), ostium primum defect, sinus venosus, and coronary sinus (least frequent). The latter two (sinus venosus and coronary sinus sub type) do not involve a developmental abnormality of the septum as such but physiologically behave as inter ASDs.\textsuperscript{3,15,21} An ostium primum ASD, (2-3\%) is the mildest form of ASD and results from the nonfusion between the septum primum and the endocardial cushions. On CT ostium, primum ASDs can be seen as septal defect located immediately posterior to the mitral valve annulus. It is commonly associated with abnormal anatomy of mitral valve, whose anterior leaflet is attached to the crest of the interventricular septum leading to abnormal left ventricle shape often angiocardio graphically identified as “goose neck deformity.” Persistent atrioventricular (AV) canal is complete variant of ostium primum defect and is associated with inlet VSD and cleft leaflets of malformed mitral and tricuspid valves. Ostium secundum ASDs (80-90\%) result from excessive apoposis of the cephalic portion of the septum primum or incomplete development of the septum secundum. On CT, ostium secundum ASDs are visualized as defects (usually larger than 1 cm) in the upper aspect of the septum behind the coronary sinus. Patent foramen ovale, not truly a septal defect per se, occur in the same position and are difficult to differentiate from ostium secundum ASD. But in general, the slit like tunnel of a patent foramen ovale will direct flow to the roof of the left atrium or to the floor of the right atrium, in contrast to the flow of ostium secundum ASDs which tends to be horizontal. Sinus venosus defects (approx 5\%) results from abnormal insertion of the superior or IVC into the right atrium or deficiency of the wall between right pulmonary veins and SVC or right atrium, producing communication between the two atria. This anomaly is thought to result from incomplete incorporation of fetal right sinus venosus into the right atrium. Superior sinus venosus defect (2-3\% of all ASD) is located at the superior cavo-atrial junction and is commonly associated with PAPVC. On CT, besides the defect, high density contrast pooling is seen in the dependent posterior portion of the left atrium (“fallen contrast sign”) which is an indirect clue to the diagnosis. Inferior sinus venosus defect is located at the junction of the IVC and right atrium. It may be associated with anomalous drainage of the right inferior pulmonary vein into the intrapericardial segment of the IVC or right atrium. Unroofed coronary sinus (least common) is said to be present when there is abnormal lack of septation between the posterior wall of the left atrium and the “unroofed” coronary sinus, resulting in abnormal shunt of venous blood into the left atrium. The coronary sinus communicates with the left atrium before draining into the right atrium. Persistent left SVC is a frequent association owing to the failure of degeneration of the left anterior cardinal vein. CT reveals a poor opacification of the coronary sinus upstream from the defect and equalization relative to the enhanced left atrium at the unroofed portion.

Atresia of the coronary sinus is a rare cardiac anomaly also associated with ASD and persistent left SVC.\textsuperscript{22}

VSD

Like with the imaging of ASD, adequate opacification of right heart is obtained by biphasic protocol (initial contrast bolus at a rate of 4-7 ml/s, followed by 40-50 ml of saline).\textsuperscript{23} Congenital VSD is the most common congenital heart anomaly and can result from maldevelopment of any of the three precursors to septum formation i.e., primitive interventricular septum (forming muscular part), caudal part of bulbar septum (forming crista of right ventricle), and down growth from endocardial cushion (forming the membranous part). A VSD is said to be small when the diameter area is less than that of the aortic annulus and large when it is more than that of the aortic annulus. It can be classified according to their location in the septum as perimembranous, muscular, subarterial, or inflow.\textsuperscript{3,15,24}

Membranous VSD (75-80\%), also known as subaortic, infracristal, perimembranous, or paramembranous occur in the upper aspect of the septum, located usually below the crista supraventricularis on right ventricle side and below the right and noncoronary cusp on the left ventricle side.
Anterior membranous VSD can be associated with TOF. Posterior membranous VSD is uncommon. Approximately one third of isolated perimembranous VSDs close spontaneously by apposition of the septal leaflet of the tricuspid valve or prolapse of right or noncoronary aortic cusp into the defect. This mechanism of spontaneous defect closure is frequently marked by ventricular septal aneurism, consists of a portion of the septal leaflet of the tricuspid valve. On CT, these defects are seen as a linear contrast filling jet crossing through the septal defect and entering the right ventricle which is washed out by biphasic contrast protocol. Gerbode-type defects are due to maldevelopment of AV cushion and lead to shunt from the left ventricle to the right atrium. They can be further classified in relation to the tricuspid valve annulus as infravalvular (involving interventricular portion of the membranous septum and fenestrations of the tricuspid septal leaflet resulting in the left ventricle to right atrium shunting) or supravalvular (involving AV portion of the membranous septum resulting in a direct communication). Muscular VSD (Maladie De Roger) consist of one or more defect in the muscular portion of the interventricular septum. The presence of multiple defects is also described as the “Swiss cheese VSD.” On CT high attenuation contrast filling tracks are seen traversing the defects. Although it can be confused with entities such as the besian sinuses, deep crypts, and coarse trabeculations, presence of contrast material entering the washed out right ventricle procures the diagnosis of muscular VSD with reasonable certainty. These VSD often regress spontaneously. VSDs located below the semilunar valve and above the crista supraventricularis are known as subarterial, outlet, supracristal, subpulmonary, infundibular, doubly committed, or conosetal VSDs. These defects often cause aortic regurgitation and prolapse of the right sinus of valsalva which may reduce or obliterate the septal defect. Inflow VSDs always occur in association with endocardial cushion defects and are associated with trisomy 21. These defects are bound by the tricuspid valve annulus and extend to the muscular septum and variably to the membranous septum. An aneurysm of membranous ventricular septum is an uncommon congenital defect associated with VSDs which can lead to systemic emboli, endocarditis, cardiac arrhythmias, left or right ventricular outflow tract obstruction and right-to-left shunts secondary to aneurismal ruptures. MDCT with 3D reconstructional images can detect these anomalies with increased accuracy.\textsuperscript{13}

**Ebstein’s anomaly**

It is an uncommon congenital anomaly characterized by apical displacement of large and redundant septal and posterior tricuspid valve leaflets with a variable degree of malformation and mobility of the anterior leaflet depending on degree of tethering to the right ventricle. The proximal right ventricle is “atrialized” as it lies on the atrial side of the tricuspid valve.\textsuperscript{8}

**Uhl’s anomaly (parchment right ventricle)**

This is an extremely rare anomaly with near complete absence of right ventricle myocardium. Right endocardium and pericardium are in contact. Right ventricle is massively enlarged and nonsystolic. This condition usually results in death in infancy or early childhood.\textsuperscript{15}

**Tricuspid atresia**

Maldevelopment of right AV cushion leads to malformed tricuspid valve either characterized by a fatty bar between the right atrium and the right ventricle or less commonly atretic valve. It is always associated with ASD. Other frequent associations are pulmonic stenosis, VSD and transposition of great arteries (TGA).\textsuperscript{3}

**CONNECTION ABNORMALITIES**

**TGA**

When a great vessel arises from inappropriate ventricle, it is said to be transposed. In TGA, aorta arises from the right ventricle and pulmonary artery arises from the left. The common type of transposition of the great arteries is a d-TGA which consists of AV concordance and ventriculoarterial discordance. It is often denoted as “S,D,D,” (S for visceroatrial situs solitus, the D for a d-loop, i.e., morphologic right ventricle on the right side and the morphologic left ventricle on the left side, and the second D stands for d-malposition of the aortic valve in relation to the pulmonary valve). In d-malposition, the aortic valve is situated to the right of and anterior to the pulmonary valve, and the great arteries are parallel instead of crossing as they do in the normal heart. Multiple communications exist between systemic and pulmonary circulations to sustain survival such as PDA, ASD, and VSD. Pulmonary stenosis is also a frequent association. Another relatively rare type of TGA is L-TGA, which consists of AV discordance and ventriculoarterial discordance. It is assigned the annotation “S,L,L,” (S for situs solitus, L for an l-loop, i.e., morphologic right ventricle positioned to the left of the morphologic left ventricle, and the second L for l-malposition of the aortic valve in relation to the pulmonary valve). The two outflow tracts are parallel, and the ventricular septum lies in a more anteroposterior...
position. The blood flow to the circulation is corrected as pulmonary vein supplies left atrium, via right ventricle to aorta and hence it is given the name “congenitally corrected TGA.” Recently, hybrid imaging techniques such as single-photon emission tomography-CT and positron emission tomography-CT with 15 O-n-ammonia has been used for patients with abnormal anatomy of the heart, including ccTGA.

**DORV**

DORV is a frequent association with many cardiac anomalies and said to be present when both aorta and pulmonary artery predominantly (>50%) originate from the right ventricle. Pulmonary artery may arise completely from the right ventricle or have biventricular origin, and aorta arises from the right ventricle.

Loss of continuity between the mitral valve and the neighboring semilunar valve is prerequisite for the diagnosis. A VSD is always present and may be subaortic (50%), subpulmonary (30%), noncommitted, or doubly committed.

- Subaortic VSD causes predominant flow from the left ventricle to the aorta, similar to TOF and is associated with pulmonary stenosis
- A subpulmonary VSD found in Taussig–Bing anomaly, causes preferential flow from the left ventricle to pulmonary artery with consequential overcirculation and the hemodynamics resembles transposition of the great arteries
- VSD beneath origin of both great vessels is doubly committed VSD
- VSD away from both origins is non-committed VSD
- Other variants include subaortic VSD without pulmonary stenosis or univentricular heart, consisting DORV with mitral atresia, an unbalanced AV canal, or severe hypoplasia of one of the ventricular sinuses.

Besides the relationship between the VSD and the great arteries, the degree of outflow obstruction, the size of the ventricles and the anatomy of the AV valves should be assessed to assist the surgical management of DORV. ECG-gated CT might be beneficial for better delineation of the position of the VSD.

Double outlet left ventricle is very rare.

**Isomerism**

Isomerism refers to symmetric bronchial, pulmonary, and atrial morphology. In right atrial isomerism, bilateral atria have a right atrial morphology identified on CT by a triangular appendage with a wide opening characterizing the morphologic right atrial appendage, both main bronchi are symmetrically short, and the upper lobe bronchi are bilaterally eparterial. Since there is no left atrium, total anomalous pulmonary venous drainage is understandably common. The liver is midline, and stomach shows variable position. Spleen is absent, and hence, this complex is also reasonably known as asplenia syndrome (Ivemark syndrome). Another finding in right isomerism is juxtaposition of the IVC and the abdominal aorta. In left atrial isomerism, bilateral atria have a finger-like appendage with a narrow opening characterizing the morphologic left atrial appendage, both main bronchi are symmetrically long, and the upper lobe bronchi are bilaterally hyparterial. Interruption of the IVC is typical and highly specific for this condition. Pulmonary veins may drain directly into nearest atrium. Spleen is usually replaced by multiple splenunculi on both sides of the abdomen, giving prompting the use of polysplenia syndrome for this disease complex. Liver is midline, and stomach can be anywhere.

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